Clinical features of children and adults with a muscular dystrophy using powered indoor/outdoor wheelchairs: disease features, comorbidities and complications of disability.

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*** Stanmore Specialist Wheelchair Service has now been disbanded**

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Implications for Rehabilitation

- **Powered wheelchairs have therapeutic benefits in managing muscular dystrophy pain and weakness**
- **The use of specialised seating needs careful consideration in supporting progressive muscle weakness and the management of scoliosis.**
- **Pain, discomfort, pressure risk and muscle fatigue may be reduced by use of tilt-in-space.**

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Abstract

Purpose: To describe the clinical features of electric powered indoor/outdoor wheelchair users with a muscular dystrophy, likely to influence optimal prescription; reflecting features of muscular dystrophies, conditions secondary to disability and comorbidities impacting on equipment provision. Methods: cross-sectional retrospective case note review of recipients of electric powered indoor/outdoor wheelchairs provided by a specialist regional wheelchair service. Data on demography, diagnostic/clinical and wheelchair prescription were systematically extracted. Results: Fifty-one men and 14 women, mean age 23.7 (range 10-67, sd 12.95) years, were studied. Forty had Duchenne muscular dystrophy, 22 had other forms of muscular dystrophy and three were unclassified. Twenty-seven were aged under 19. Notable clinical features included problematic pain (10), cardiomyopathy (5) and ventilatory failure (4). Features related to disability were (kypho)scoliosis (20) and oedema/cellulitis (3) whilst comorbidities included back pain (5). Comparison of younger with older users revealed younger users had more features of muscular dystrophy affecting electric powered chair provision (56%) whilst older users had more comorbidity (37%). Tilt-in-space was prescribed for 81% of users, specialised seating for 55% and complex controls for 16%. Conclusions: Muscular dystrophy users were prescribed electric powered indoor/outdoor chairs with many additional features reflecting the consequences of profound muscle weakness. In addition to

facilitating independence and participation, electric powered indoor/outdoor chairs have major therapeutic benefits.

Introduction

Muscular dystrophy, including myotonic dystrophy, is a term covering a set of chronic, hereditary and progressive diseases involving degeneration of muscle cells and leading to successive wasting of the muscles including the respiratory muscles.[1] Limb girdle muscular dystrophy is now recognised as a heterogeneous group of myopathies that vary in severity and may affect persons of all ages from childhood through adulthood.[2] Life expectancy has improved markedly in recent years and Duchenne muscular dystrophy is now considered a disease of adulthood.[3]

The muscular dystrophies are rare diseases, with a 'combined prevalence between 19.8 and 25.1 per 100,000 person years'.[4] The prevalence of the commonest types are fascioscapulohumeral dystrophy (3.2-4.6 per 100,000), myotonic dystrophy (0.5-18.1 per 100,000) and Duchenne muscular dystrophy (1.7–4.2 per 100,000).[4] All those with Duchenne muscular dystrophy will become wheelchair reliant in childhood, usually by the age of 12 years unless treated.[5] The United Kingdom (UK) Registry of fascioscapulohumeral dystrophy has reported that about 40% of those registered used wheelchairs at least part-time.[6] The mean age of achieving wheelchair reliance for those with Becker muscular dystrophy was 50 years.[7] Use of powered wheelchairs or powered mobility has been reported from 7% for those with Duchenne muscular dystrophy [8] to 31% of individuals with neuromuscular disease.[9]The

transition of both health and social support as children transfer to adult services at the age of 19 years is recognised as problematic [10-11] with inadequate adult health provision for those ageing with childhood deteriorating conditions.[12]

Powered mobility is now becoming more important as its value is increasingly accepted not only as a means of improving mobility and independence [13-14], increasing quality of life for those with muscular dystrophies,[15] potential savings in social costs [16] but also because of its direct therapeutic effects.[17-18] Guidelines for those with limb girdle muscular dystrophies recommend the 'prescription of assistive devices that are adapted specifically for the patient's deficiencies' [2] and guidelines for those with Duchenne muscular dystrophy recommend assistive technology and manual/powered wheelchairs as part of rehabilitation management.[14] The emphasis has therefore shifted from powered wheelchairs being aids for mobility to being facilitators of participation and occupation. Additionally, it is acknowledged that powered wheelchairs are fundamental in optimising medical management for those with long-term disabling conditions.[18]

While powered wheelchairs are used to increase function and mobility their provision is equally important for minimising discomfort and postural abnormalities.[19] It is acknowledged that, as people with Duchenne muscular dystrophy grow older, they acquire a wide range of clinical complications that need management,[20] often through specialised seating (adaptive seating) on the powered wheelchair base. The major clinical issues relate to orthopaedic complications including fractures, (kypho)scoliosis, cardiopulmonary involvement [20] and pain.[9,21-22]

Postural problems such as a scoliosis are known to develop when individuals are still ambulant.[23] Specialised seating can provide appropriate postural support but this may conflict with the desire of the chair user (hereafter described as the 'user') to keep mobile within the chair [21] which may challenge a wheelchair provider. This challenge is compounded during the years of growth [21] and the curve may progress even after cessation of growth,[24] associated with the increasing muscular weakness. Regular monitoring and careful management of the spinal complications of muscular dystrophy is recommended to maintain optimal posture and cardiopulmonary function.[2]

The progressive muscular weakness characteristic of the muscular dystrophies determines the development of a range of clinical manifestations which depend on the disease trajectories of the different types of muscular dystrophy. Many of the common features of muscular dystrophy have been documented.[2,14,25] Individuals may also develop health conditions associated with long-term disability. These features of disability have been reported in other powered wheelchair users.[17,26] In addition, individuals may be diagnosed with other conditions unrelated to the muscular dystrophy, comorbidities, although they may reflect age-related health issues.[27] Collectively, these have been referred to as 'Associated Clinical Features' (ACFs) [17,26] as it is acknowledged that some ambiguity exists due to lack of research into the development of conditions across the lifespan of disease.

The aims of this study are to describe the clinical features of electric powered indoor/outdoor chair users with muscular dystrophy and to explore the complexities of comorbidities, features of muscular dystrophy and conditions secondary to disability that impact on powered wheelchair provision and clinical

management. A further aim is to compare the clinical features and electric powered indoor/outdoor chair provision of those under the age of 19 years with those aged 19 or over.

Methods

This is a cross-sectional study of a clinic population and was approved by the National Research Ethics Service as a service evaluation.

The participants

The potential participants for this study were wheelchair users with a muscular dystrophy referred from locality based wheelchair services to a specialist wheelchair service serving a mixed population from inner city, suburban and rural areas of approximately 3.1 million. Following assessment of suitability for an electric powered indoor/outdoor chair, those who fulfilled eligibility criteria [28] were included. Details of the specialist wheelchair service and the full assessment for electric powered indoor/outdoor chair eligibility have been described elsewhere.[26] In essence, those unable to self-propel and who were able to take advantage of using a powered chair indoors and outdoors safely were included. The study participants had been provided with an electric powered indoor/outdoor chair and were currently using it.

Procedures

Data consisting of demographic data, clinical issues and wheelchair factors were entered into electronic records and reviewed between June 2007 and September 2008 by a consultant physician in rehabilitation medicine. They were systematically extracted and entered into a computer database. Average time

from the date of initial assessment for an electric powered indoor/outdoor chair in clinic to the case note review was 5.28 (range 0-10.7, sd 3.23) years. Further data were entered from clinical notes (charts) and anonymised.

Demographic profiles consisted of age at initial assessment and gender. Where the database held no precise diagnostic information relating to the type of muscular dystrophy, it was classified as 'unknown'.

Clinical profiles perceived to be relevant to the electric powered indoor/outdoor chair prescription included comorbidities, complications relating to muscular dystrophy or disability. Due to the ambiguity in which some clinical features may reflect either the muscular dystrophy itself, co-morbidity or a complication of disability, they were grouped as ACFs when referred to collectively.[17] Users requiring further clinical management were referred to their family doctor (primary care doctor).

Pain is a major problem for many with a muscular dystrophy.[14] Users with pain requiring further investigation/management, or influencing the electric powered indoor/outdoor chair prescription were recorded as 'problematic pain'.[17,26]

Features noted by others as being disability related were also included. They were spinal deformities,[29-30] osteoporosis, [5,31] pelvic obliquity,[29] contractures, [2,5,32] thrombo-embolism,[33] dependent oedema [18,34] and pressure sores.[21] Conditions classified as comorbidities were those considered unrelated to muscular dystrophy.

Wheelchair factors included information about specialised seating, defined as that which is 'required by those who need a wheelchair for their mobility but due to postural instability or deformity need extra support in order to function'[35,p.7]. Other data included tilt-in-space, cushions, complex controls, chair mounted ventilation and use of the National Health Service Voucher Scheme.[36]

Methods of analysis

This was a secondary data analysis of a sub-group of electric powered indoor/outdoor chair users from a cohort reported elsewhere.[37] Descriptive statistics were used to analyse demographic, clinical and wheelchair data.

Results

Sixty five users mean age 23.7 (range 10-67, sd 12.95) years (table1), comprised 51men mean age 22.6 (range 10-67, sd 12.9) years and 14 women mean age 27.7 (range 10-49, sd 12.9) years. The dominant group of users had Duchenne muscular dystrophy and this group were the youngest with a mean age of 18 years (table 1).Twenty seven users were aged under 19 years, the youngest aged 10, whilst 38 users were aged 19 and older, the oldest aged 67 (table 2).

Suggest insert table 1 and table 2 about here.

Associated clinical features

Twenty five users had no ACFs. Nineteen had one ACF, 14 had two ACFs whilst seven had more than two ACFs. Of the 40 users with Duchenne muscular dystrophy, 27 had at least one ACF.

We noted 10 comorbidities reported in 18 users, the most common being back pain in five users. Hypertension and ischaemic heart disease were noted in users aged 54, 57 and 60. Two users, both with congenital muscular dystrophy experienced cerebrovascular events; one 33 year old had a stroke and the other aged 13 had an intracranial bleed. There were nine features of muscular dystrophy reported by 26 users with problematic pain being the most frequent reported by 10 users. Scoliosis was the most frequently noted of the six complications of disability (n=20; 10 younger and 10 older users) (table 3). No reports were noted for thromboembolism.

Comparison of those under 19 years with older users revealed that the younger group had more features of muscular dystrophy affecting electric powered indoor/outdoor chair provision (56%) whilst the older users had more comorbidity (37%). The complications of disability were proportionately similar for younger (44%) and older (42%) users (table 2). Four users had ventilatory failure, mean age 22 years, three with Duchenne muscular dystrophy and one with congenital muscular dystrophy and required oxygen cylinders to be accommodated by the electric powered indoor/outdoor chair. Two of these users had tilt-in-space and three had specialised seating. All four were provided with tray-mounted complex controls.

Thirty-seven users were noted to have clinical issues that needed further attention from their family doctor (primary care team). The majority of these

referrals related to medical issues, about half of which concerned the need for pain management.

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Wheelchair features

Tilt-in-space was provided to 48 (81% of those with known data) users (table 1). Eighty five percent of younger users were provided with tilt-in-space, compared with 66% of older users (table 2). Just over half of the cohort was provided with specialised seating (table1) with 74% of younger users compared with 42% of older users having this feature (table 2). Sixty five percent of users with Duchenne muscular dystrophy were prescribed specialised seating compared to 40% of other muscular dystrophy users. Thirty one users were provided with both specialised seating and tilt-in-space although two users with specialised seating had unknown tilt-in-space provision.

Contoured systems were provided to 12 users: carved foam (4), Scott seating systems (4), matrix (3) and moulded seat insert. The majority of the remainder had pressure relieving cushions, mainly Jay 2 cushion (14), Qbitus (10), Vicaire (4) and RoHo (3) and Other (n=7). Fifteen were provided with a standard wheelchair cushion.

Complex controls were provided to 17 users of which 14 were tray mounted, seven were non-standard and six had interfacing issues with other assistive technology devices. A further three users were referred for environmental control units.

The voucher scheme was used by three users/parents. One 11y old boy with limb girdle muscular dystrophy provided with a 'riser' function in partnership with the charity 'WhizzKidz'; another 12y old boy with Duchenne muscular dystrophy provided with a 'lying-to-stand function' in partnership with WhizzKidz; and a 12y old boy with Duchenne muscular dystrophy whose parents were very unhappy with the service provided and preferred the individual responsibility for maintenance of the electric powered indoor/outdoor chair.

Discussion

Extensive follow-ups of electric powered indoor/outdoor chair users with a muscular dystrophy are rare. This report of a UK group spans an average of 5.3 years of electric powered indoor/outdoor chair use by individuals with muscular dystrophy. Our group of users are atypical when compared to other muscular dystrophy populations. Firstly, they are all severely disabled, unable to walk, and due to the strict eligibility criteria for electric powered indoor/outdoor chair provision,[28] those with poorly controlled epilepsy, learning disability and cognitive dysfunction are under-represented. Some of these users have been reported previously at a two-year follow-up.[21]

These muscular dystrophy electric powered indoor/outdoor chair users were on average older than most other muscular dystrophy populations as age was not a criterion for selection. However, our youngest user was aged 10 years as might be expected from the likely age of loss of walking in Duchenne muscular dystrophy [38-39] and is comparable to those reported by Engle et al in a mixed population of users with both muscular dystrophy and myotonic dystrophy.[9]

Studies that include both children and adults of all ages are rare, although Pangalila has reported those with Duchenne muscular dystrophy aged up to 44.[40] Those with Becker muscular dystrophy and fascioscapulohumeral dystrophy become wheelchair dependent when older as reflected in our data and that of Wood et al.[6]

Clinical features of Muscular Dystrophies

The progressive muscular weakness characteristic of muscular dystrophies results in the decline of functional ambulation at which point powered mobility is advocated.[14] Our study indicates that the loss of ambulation can occur across the age range depending on the type of muscular dystrophy. The reasons for provision of electric powered indoor/outdoor chairs not only relate to mobility factors but also to the expectations of individual's community participation at different stages of their life.[37] The needs of younger users, in this study mainly those with Duchenne muscular dystrophy, include facilitating engagement in school and play while at the same time accommodating growth and anticipated disease progression. Older users, in this study mainly those with other forms of muscular dystrophy, have needs relating to families, work and active retirement. The independence provided by an electric powered indoor/outdoor chair facilitates psychological and social development for younger users' [41] and community living for older ones.[42]

In order to achieve this, electric powered indoor/outdoor chair prescription with appropriate additional features such as specialised seating, should be used to reduce the impact of symptoms and maintain or improve residual function. The findings of this study bring together the main technical considerations of electric

powered indoor/outdoor chair provision with clinical information relating to muscular dystrophies and severe disability.

Pain is a recognised feature of Duchenne muscular dystrophy with higher levels reported for those still mobile as compared to wheelchair users.[43] It is a significant problem for young people with Duchenne muscular dystrophy affecting most parts of the body and interfering with several life domains.[9] Engle's patients' reported the 'pain as negatively affecting many aspects of youth and adolescent life' [9] and also that some had severe pain which is consistent with our findings of problematic pain. Pain was also reported in adult patients with Duchenne muscular dystrophy but appeared not to interfere greatly with activities.[40] Back pain, together with leg and neck pain, are reported as common sites of pain in neuromuscular disorders.[9] This is consistent with our findings and indicates the need for adjustment and review of seating [21] which in this study showed that specialised seating was required by the majority of users, especially those under 19 years of age.

The incidence of neck pain was extremely low with only one user reported with this problem. The likely reason was that neck restraints were provided as standard to all our users. This is particularly important for those with progressive weakness and, in tandem with using tilt-in-space, provides appropriate support for the head and neck. This is an important contribution to their rehabilitation through facilitating head control. Provision of tilt-in-space also makes an important contribution to pain management as it allows the user to change position as frequently as they wish in order to minimise the pain and discomfort associated with prolonged sitting. Our results show a high proportion

of all muscular dystrophy electric powered indoor/outdoor chair users being provided with tilt-in-space.

Our four users with ventilatory support all had complex control systems as has been reported previously.[44] Cardiomyopathy and chronic respiratory insufficiency are universal in the later stages of Duchenne muscular dystrophy.[5] It is debateable whether respiratory insufficiency is a consequence of muscular dystrophy progressive weakness or due to chest wall and spinal deformities reducing respiratory capacity.[45] In this cohort, specialised seating systems were used therapeutically to facilitate reduction of spinal deformity during growth, although after growth seating accommodated any spinal deformity. A balance must be sought between stability and flexibility to allow those who use a 'row-a-boat' breathing to have sufficient movement in their chairs.[46]

Comorbidity

Reports of comorbidities in the muscular dystrophies are rare. However, the 'prevention and management of comorbidities' is important.[2,p.1461] In common with this study, asthma has been reported as a comorbidity in a sample of youths [9] as has an increased risk of bone fractures in Duchenne muscular dystrophy.[5,47] The one user we noted with multiple fractures of both distal femurs and proximal tibiae had tipped out of his wheelchair, a known risk for electric powered indoor/outdoor chair users.[28] Also consistent with previous studies,[48] we had two users with epilepsy – one noted to be temporal lobe, the other unspecified. This may be unrepresentative as some potential users with

epilepsy will not fulfil the eligibility criteria [28] due to inadequate epilepsy control. We have noted epilepsy as a comorbidity although recent research may indicate it is part of the spectrum of Duchenne muscular dystrophy.[49]

Comorbidities such as hypertension and ischaemic heart disease may be considered due to the ageing process, possibly related to the sedentary lifestyle consequent to wheelchair reliance. The cerebrovascular events noted for two electric powered indoor/outdoor chair users with congenital muscular dystrophy appear not to have been reported previously, but it is speculated that they may be due to cortical abnormalities noted in this condition.[50]

Disability features

One of the greatest challenges for wheelchair prescribers and those with muscular dystrophies is the development of a scoliosis. [24,51-52] It has been reported that the magnitude of the curve was predicted by the length of time using a wheelchair. [53] We found 20 (31%) users with a scoliosis that required addressing through the electric powered indoor/outdoor chair provision. Seeking an appropriate balance between stabilising the spine and retaining upper limb and trunk flexibility in the system is complex and the views of the user and their family's priorities may produce tensions that need resolution by the seating team.[26] For example, many of the users also used their manual backup chair to extend their social life beyond the reach of their powered chair.[41] This creates extra physical strain for the parents of these young users when they have to transfer the seating system from their electric powered indoor/outdoor chair to their manual chair. Postural issues are critical in this group and are greatly

complicated by the influence of growth often needing adjustments to the electric powered indoor/outdoor chair within one year of prescription.[21]

Dependent oedema is a well established complication of prolonged sitting [18] and has been noted previously in other electric powered indoor/outdoor chair user groups [17,26] but may also be due to electrolyte imbalance in Duchenne muscular dystrophy.[54]

Pressure problems are well recognised for wheelchair dependent wheelchair (not necessarily powered wheelchair) users with Duchenne muscular dystrophy.[55] Pressure problems appear to be better controlled now than when reported from this service previously.[21] However, the presence of two pressure sores suggests that further measures need to be taken to ensure adequate protection against pressure.

Wheelchair issues

In contrast to other electric powered indoor/outdoor chair user groups, [17,26] these users with muscular dystrophies had a higher frequency of provision of SS and TIS which was particularly found for those under 19 years of age with almost half the cohort being provided with both specialised seating and tilt-in-space. We also found that the users with Duchenne muscular dystrophy were most frequently provided with specialised seating. This indicates both the severity of the disability, the likelihood of its progression and the complexity of meeting individuals' needs through the rehabilitation process.

The value of tilt-in-space has been noted previously for those with muscular dystrophy. [21,34] It has been recommended as good practice in the management of muscular dystrophies for reducing the impact of gravity on a scoliosis, pressure redistribution, increasing comfort and pain relief, reducing fatigue, increasing independence for the chair user to change position and assisting carers in moving/handling in the management of personal care.[34] For this cohort of electric powered indoor/outdoor chair users with muscular dystrophy, these issues were considered across the age range to include chair use at home, in school, college/university, the workplace and the outdoor environment.

Specialised seating is primarily provided to improve sitting posture, but is also reported to facilitate small improvement in upper limb function,[51] both of which are important for facilitating function and participation. For users who are children or adolescents, specialised seating may require change or adaptation to accommodate growth.[21] Those needing the most support in this study (12 users) were provided with individually fitted specialised seating. These are likely to be the most severely impaired (e.g. through a scoliosis) and vulnerable individuals with muscular dystrophies maintained in the community. Specialised seating was provided for a number of other reasons, the most important being pressure redistribution, prolongation of sitting time, preventing contractures and providing the additional support needed to negotiate ramps and slopes.[34]

Trays can be used to provide additional support for the upper limbs when arm rests prove insufficient.[34] This is particularly important where muscle fatigue/weakness limits the residual functions of the hands to control the electric powered indoor/outdoor chair. Provision of a tray-mounted control box

alleviated this issue for 14 of our users, six of whom utilised the tray for interfacing with other equipment. Where the control box can be positioned centrally, a more symmetrical posture is achieved. Complex controls, when needed, included light touch systems appropriate for those with extreme muscle weakness as reported previously.[44]

For the four users who needed ventilatory support, the electric powered indoor/outdoor chair prescription would have to take into account the weight of the oxygen cylinders which are usually mounted on the back of the chair. The engineers would allow for the use of tilt in making their prescription. It is not surprising that those whose cardio-respiratory weakness required ventilatory support also had such severe peripheral weakness that tray-mounted complex controls were needed.

Health-related quality of life is severely affected by Duchenne muscular dystrophy [56] and the consequent socioeconomic costs of direct, indirect and informal care costs are substantial.[56] The complexity of meeting the requirements of those with muscular dystrophy who need powered mobility is reflected in the electric powered indoor/outdoor chair prescription. Thus the proportion of those with muscular dystrophy being provided with tilt-in-space, tilt-in-space and specialised seating, and complex controls is greater than for those with multiple sclerosis,[17] rare diseases,[26] spinal cord injuries [57] and cerebral palsy [58]. The need for these extra features increases the capital cost of providing the electric powered indoor/outdoor chair, and also the need for extra time from the highly skilled team of rehabilitation professionals giving the assessment, provision and maintenance of the chair.

Strengths and limitations

This is one of the few studies with a five year period of follow-up that addresses the needs of both children and adults severely affected with a muscular dystrophy who use powered wheelchairs. A strength of this study was that all users were seen by the same multiprofessional team who were very experienced in the provision of powered wheelchairs and therefore the clinical decisions made were consistent and in accordance to service protocols.

Our electric powered indoor/outdoor chair users with muscular dystrophy were predominately those with Duchenne muscular dystrophy. Therefore, it is difficult to comment further on the more specific needs of those with other types of muscular dystrophy. In addition, we lacked precise diagnostic data regarding the type of muscular dystrophy in three users. We acknowledge missing data on the presence or absence of tilt-in-space in six users.

The service records did not gather data on the use of elevating leg rests or recline features of the electric powered indoor/outdoor chair provided and this is regrettable as these features have major therapeutic benefits, but at additional costs.

The nature of the provision of electric powered indoor/outdoor chairs in the UK at the time of the study meant that electric powered indoor/outdoor chair s were provided to very few children below secondary school age (10-11 years) and thus the undoubted benefits of powered mobility for younger children were not assessed.

We recognise that the presence of ACFs is likely to be under-reported as data was obtained from referral letters and patient histories. However, such data is more likely to be objective than purely patient self-report surveys. Further prospective studies are recommended.

This study did not include those who used scooters or had purchased their powered wheelchairs privately or with charitable funding (mostly available for children only).

Conclusions

In addition to facilitating independence and participation for those with a muscular dystrophy, powered wheelchairs have major therapeutic benefits, particularly in the management of wheelchair users' pain and weakness in addition to the more recognised benefits of pressure relief.

A (kypho)scoliosis, problematic pain and ventilatory failure were the clinical features most likely to influence the electric powered indoor/outdoor chair prescription. Frequent use of specialised seating and tilt-in-space were utilised to facilitate management of these clinical complications, whilst accommodation of the ventilatory equipment required adjustments to the electric powered indoor/outdoor chair.

The use of powered tilt-in-space, specialised seating and complex controls was greater than for any other group of electric powered indoor/outdoor chair users studied to date and this reflects the complexity of meeting the needs of those with profound peripheral muscle weakness. This utilisation was greater for the children than for those aged 19 years or older and is likely to reflect not only

growth issues but also the rapid deterioration of those with Duchenne muscular dystrophy and some other congenital forms of muscular dystrophy. The need for a high use of these electric powered indoor/outdoor chair features will have cost implications for neuromuscular service providers.

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Reference List

(1) Ahistrom G, Lindvall B, Wenneberg S, Gunnarsson LG. A comprehensive rehabilitation programme tailored to the needs of adults with muscular dystrophy. Clin Rehabil 2006;20:132-41.

(2) Narayanaswami P, Weiss M, Selcen D et al. Evidence-based guideline summary: diagnosis and treatment of limb-girdle and distal dystrophies: report of the guideline development subcommittee of the American Academy of Neurology and the practice issues review panel of the American Association of Neuromuscular & Electrodiagnostic Medicine. Neurol 2014;83(16):1453-63.

(3) Passamano L, Taglia A, Palladino A et al. Improvement of survival in Duchenne Muscular Dystrophy: Retrospective analysis of 835 patients. Acta Myol 2012;31(2):121-25.

(4) Theadom A, Rodrigues M, Roxburgh R, Balalla S, Higgins C, Bhattacharjee R et al. Prevalence of muscular dystrophies: a systematic literature review. Neuroepid 2014;43(3-4):259-68.

(5) Yiu EM, Kornberg AJ. Duchenne muscular dystrophy. J Paediat Child Health 2015;51(8):759-64.

(6) Wood L, Evangelista T, Williams M, Smith D, Norwood F, Rogers M et al. UK patient registry for facioscapulohumeral muscular dystrophy (FSHD). Neuromusc Disord 2015;25:S36.

(7) Van Den Bergen JC, Wokke BHA, Van Damme P, Van Der Kooi AJ, De Bleecker J, De Jonghe P et al. Development of a disease severity scale for Becker muscular dystrophy. Neuromusc Disord 2014;24(9-10):795.

(8) Hilbert JE, Kissel JT, Luebbe EA, Martens WB, McDermott MP, Sanders DB et al. If you build a rare disease registry, will they enroll and will they use it? Methods and data from the National Registry of Myotonic Dystrophy (DM) and Facioscapulohumeral Muscular Dystrophy (FSHD). Contemporary Clin Trials 2012;33(2):302-11.

(9) Engel JM, Kartin D, Carter GT, Jensen MP, Jaffe KM. Pain in youths with neuromuscular disease. Am J Hospice Pall Care 2009;26(5):405-12.

(10) Rodger S, Steffensen BF, Lochmuller H. Transition from childhood to adulthood in Duchenne muscular dystrophy (DMD). Orphanet J Rare Dis 2012;suppl 2:7.

(11) McAdam LC, Mah J, Biggar WD. Becoming an adult with Duchenne muscular dystrophy in Canada. Neuromusc Disord 2014;24(9-10):855.

(12) Rodger S, Woods KL, Bladen CL et al. Care provision for adults with Duchenne muscular dystrophy in the UK: Compliance with international consensus care guidelines. Neuromusc Disord 2015;25:S12.

(13) Davies A, De Souza LH, Frank AO. Changes in the quality of life in severely disabled people following provision of powered indoor/outdoor chairs. Disabil Rehabil 2003;25(6):286-90.

(14) Bushby K, Finkel R, Birnkrant DJ et al. Diagnosis and management of Duchenne muscular dystrophy, part 2: implementation of multidisciplinary care. The Lancet Neurol 2010;9(2):177-89.

(15) Pousada García T, Groba Gonzílez B, Nieto Rivero L et al. Exploring the Psychosocial Impact of Wheelchair and Contextual Factors on Quality of Life of People with Neuromuscular Disorders. Assist Technol : the official journal of RESNA 2015;27(4):246-56.

(16) Salatino C, Andrich R, Converti RM, Saruggia M. An observational study of powered wheelchair provision in Italy. Assist Technol : the official journal of RESNA 2016;28(1):41-52.

(17) De Souza LH, Frank AO. Problematic clinical features of powered wheelchair users with severely disabling multiple sclerosis. Disabil Rehabil 2015;37(11):990-96.

(18) Dicianno BE, Lieberman J, Schmeler M et al. RESNA position on the application of tilt, recline, and elevating leg rests for wheelchairs: 2015 current state of the literature. Arlington, VA, USA: RESNA; 2015.

(19) Mannlein J, Pangilinan PH. Wheelchair seating for children with Duchenne Muscular Dystrophy. J Ped Rehabil Med 2008;1(3):225-35.

(20) Parker AE, Robb SA, Chambers J, Davidson AC, Evans K, O'Dowd J et al. Analysis of an adult Duchenne muscular dystrophy population. Quart J Med 2005;98(10):729-36.

(21) Richardson M, Frank AO. Electric powered wheelchairs for those with muscular dystrophy: problems of posture, pain and deformity. Disabil Rehabil Assist Technol 2009;4(3):181-88.

(22) Smith AE, McMullen K, Jensen MP, Carter GT, Molton IR. Symptom burden in persons with myotonic and facioscapulohumeral muscular dystrophy. Am J Phys Med Rehabil 2014;93(5):387-95.

(23) Lord J, Behrman B, Varzos N, Cooper D, Lieberman JS, Fowler WM. Scoliosis associated with Duchenne muscular dystrophy. Arch Phys Med Rehabil 1990;71(1):13-7.

(24) Heller KD, Forst R, Forst J, Hengstler K. Scoliosis in Duchenne muscular dystrophy: aspects of orthotic treatment. Prosthet Orthot Int 1997;21(3):202-09.

(25) Narayanaswarmi P, Weiss M, Selsen D et al. Evidence-based guideline summary: evaluation, diagnosis, and management of congenital muscular dystrophy: Report of the Guideline Development Subcommittee of the American Academy of Neurology and the Practice Issues Review Panel of the American Association of Neuromuscular & Electrodiagnostic Medicine. Neurol 2015;84(13):1369-78.

(26) De Souza LH, Frank AO. Rare diseases: matching wheelchair users with rare metabolic, neuromuscular or neurological disorders to electric

powered indoor/outdoor wheelchairs (EPIOCs). Disabil Rehabil 2015;38:1547-56.

(27) Naidoo V, Putnam M, Spindel A. Key focal areas for bridging the fields of aging and disability: findings from the growing older with a disability conference. Int J Integrated Care 2012;12:e201.

(28) Frank AO, Ward JH, Orwell NJ, McCullagh C, Belcher M. Introduction of the new NHS Electric Powered Indoor/outdoor Chair (EPIOC) service: benefits, risks and implications for prescribers. Clin Rehabil 2000;14:665-73.

(29) Scannell BP, Yaszay B, Bartley CE, Newton PO, Mubarak SJ. Surgical Correction of Scoliosis in Patients With Duchenne Muscular Dystrophy: 30-Year Experience. J Pediatr Orthop 2016 Jan 11. [Epub ahead of print]

(30) Harvey A, Baker L, Williams K. Non-surgical prevention and management of scoliosis for children with Duchenne muscular dystrophy: what is the evidence? J Paediatr Child Health 2014;50(10):E3-E9.

(31) Tian C, Wong B, Hornung L et al. Age-specific prevalence of osteoporosis and frequency of poor bone health indices in duchenne muscular dystrophy. Neuromusc Disord 2014;24(9-10):857.

(32) Narayanaswarmi P, Weiss M, Selsen D et al. Evidence-based guideline summary: evaluation, diagnosis, and management of congenital muscular dystrophy: Report of the Guideline Development Subcommittee of the American Academy of Neurology and the Practice Issues Review Panel of the

American Association of Neuromuscular & Electrodiagnostic Medicine. Neurol 2015;84(13):1369-78.

(33) Coveney E, O'Halpin D, Fitzgerald RJ. Spontaneous deep venous thrombosis in Duchenne muscular dystrophy. Pediatr Surg Int 1993;8(4):368-70.

(34) Muscular Dystrophy Campaign. Wheelchair provision for children and adults with muscular dystrophy and other neuromuscular conditions: best practice guidelines. 2nd ed. London: Muscular Dystrophy Campaign; 2011.

(35) British Society of Rehabilitation Medicine. Specialised wheelchair seating national clinical guidelines. Report of a multidisciplinary expert group (Chair: Marks, LJ). London: British Society of Rehabilitation Medicine; 2004

(36) Frank AO, Ellis K, Yates M. Use of the voucher scheme for provision of Electric Powered Indoor/outdoor Wheelchairs (EPIOCs). Posture & Mobility 2008;23(1):17-26.

(37) Frank AO, De Souza LH. Recipients of Electric Powered Indoor/outdoor Wheelchairs (EPIOCs) provided by a National Health Service: a cross sectional study. Arch Phys Med Rehabil 2013;94:2403-9.

(38) Bushby K. Duchenne Muscular Dystrophy: medical factsheet. London, Muscular Dystrophy Campaign; 2002.

(39) Werge B, Rahbek J, Madsen A, Marquardt J, Werlauff U, Steffensen BF. Physical ability and health in a non-steroid population of 77 adult patients with Duchenne muscular dystrophy. Neuromusc Disord 2014;24(9-10):854-5.

(40) Pangalila RF, van den Bos GA, Bartels B, Bergen M, Stam HJ, Roebroeck ME. Prevalence of Fatigue, Pain, and Affective Disorders in Adults With Duchenne Muscular Dystrophy and Their Associations With Quality of Life. Arch Phys Med Rehabil 2015;96(7):1242-7.

(41) Evans S, Neophytou C, De Souza LH, Frank AO. Young people's experiences using electric powered indoor-outdoor wheelchairs (EPIOCs): potential for enhancing users' development? Disabil Rehabil 2007;19(16):1281-94.

(42) Evans S, Frank A, Neophytou C, De Souza LH. Older adults' use of, and satisfaction with, electric powered indoor /outdoor wheelchairs. Age and Ageing 2007;36(4):431-5.

(43) Douvillez B, Braillon P, Hodgkinson I, Berard C. Pain, osteopenia and body composition of 22 patients with Duchenne muscular dystrophy: A descriptive study. Ann Readapt Med Phys 2005;48(8):616-22.

(44) Pellegrini N, Guillon B, Prigent H et al. Optimization of power wheelchair control for patients with severe Duchenne muscular dystrophy. Neuromusc Disord 2004;14(5):297-300.

(45) Umayahara Y, Nakashima M, Kuwata M, Sekikawa K. Relationships between spinal deformities and chest wall deformities in patients with duchenne muscular dystrophy. Physiother (United Kingdom) 2015;101:eS1557.

(46) Yasuma F, Kato T, matsuoka Y, Konagaya M. Row-a-boat phenomenon: respiratory compensation in advanced Duchenne Muscular Dystrophy. Chest 2004;119(6):1836-39.

(47) James KA, Cunniff C, Apkon SD et al. Risk Factors for First Fractures Among Males With Duchenne or Becker Muscular Dystrophy. J Pediatr Orthop 2015;35(6):640-4 .

(48) Cuijie W, Haipo Y, Xiaona F et al. Duchenne and Becker muscular dystrophy complicated with epilepsy. Chin J Pediatr 2015;53(4):274-9.

(49) Sandu C, Tarta-Arsene O, Butoianu N, Craiu D, Iliescu C. Epilepsy in Duchene muscular dystrophy patients. Eur J Paediatr Neurol 2013;17:S134.

(50) Kang PB, Morrison L, Iannaccone ST et al. Evidence-based guideline summary: evaluation, diagnosis, and management of congenital muscular dystrophy: Report of the Guideline Development Subcommittee of the American Academy of Neurology and the Practice Issues Review Panel of the American Association of Neuromuscular & Electrodiagnostic Medicine. Neurol 2015;84(13):1369-78.

(51) Clark J, Michael S, Morrow M. Wheelchair postural support for young people with progressive neuromuscular disorders. Int J Ther Rehabil 2004;11(8):3765-73.

(52) Furderer S, Hopf C, Zollner J, Eysel P. Scoliosis and hip flexion contracture in Duchenne muscular dystrophy. Zeitschrift fur Orthopadie und Ihre Grenzgebiete 2000;138(2):131-5.

(53) Shapiro F, Zurakowski D, Bui T, Darras BT. Progression of spinal deformity in wheelchair-dependent patients with Duchenne muscular dystrophy who are not treated with steroids: Coronal plane (scoliosis) and sagittal plane (kyphosis, lordosis) deformity. Bone and Joint J 2014;96 B(1):100-5.

(54) Lehmann-Horn F, Weber MAü, Nagel AM, Meinck HM, Breitenbach S, Scharrer J et al. Rationale for treating oedema in Duchenne muscular dystrophy with eplerenone. Acta Myol 2012;31:31-9.

(55) Liu M, Mineo K, Hanayama K, Fujiwara T, Chino N. Practical problems and management of seating through the clinical stages of Duchenne's muscular dystrophy. Arch Phys Med Rehabil 2003;84(6):818-24.

(56) Landfeldt E, Lindgren P, Bell C F et al. The burden of muscular dystrophy: An international cross-sectional study. Neurol 2014;83:529-36.

(57) De Souza LH, Frank AO. Matching wheelchair users with severe spinal cord injuries to provision of electric powered indoor/outdoor wheelchairs (EPIOCs). Paper presented at: The 5th European Seating Symposium; 2016 June 13-15; Dublin, Republic of Ireland.

(58) Frank AO, De Souza LH. Problematic clinical features of children and adults with cerebral palsy who use electric powered indoor/outdoor wheelchairs: A cross-sectional study, Assistive Technology 2016, DOI:10.1080/10400435.2016.1201873

Table 1. Age, diagnosis and wheelchair provision for 65 users with a Muscular Dystrophy

Type of muscular dystrophy	No. (No. Male)	Mean age (range) (years)	Special Seating	Tilt-in-space **	Complex controls	Scoliosis	Problematic pain
Becker's	2 (2)	42 (24-60)	0	1	0	0	0
Congenital	11 (2)	23 (11-34)	7	9	3	5	1
Duchenne	40 (40)	18 (10-34)	26	31	13	14	8
Dystrophia Myotonica	1 (1)	54 *	0	1	0	0	0
Fascio-scapulo-humeral	2 (2)	45 (33-57)	0	0	0	0	0
Limb-girdle	6 (2)	36 (11-49)	2	4	1	0	1
Unknown	3 (2)	37 (10-67)	1	2	0	1	0
Total	**65 (51)**	**23.7 (10-67)**	**36**	**48**	**17**	**20**	**10**

***** **Only one user**

****** **Data unknown for 6 users**

Table 2. Clinical features and wheelchair provision for 27 users with a muscular dystrophy aged under 19 and 38 users aged 19 or over.

Age group	Under 19	19 or more	Total
No. of users	**27**	**38**	**65**
Mean age (range)	**13.8** **(10 – 18)**	**30.7** **(19 – 67)**	**23.7** **(10 – 67)**
Special Seating n (%)	**20 (74)**	**16 (42)**	**36 (55)**
Tilt-in-Space n (%) *	**23 (85)**	**25 (66)**	**48 (74)**
Complex controls n (%)	**6 (22)**	**11 (29)**	**17 (26)**
Features of muscular dystrophy n (%)	**15 (56)**	**11 (29)**	**26 (40)**
Comorbidity	**4 (15)**	**14 (37)**	**18 (28)**
Complications of disability	**12 (44)**	**16 (42)**	**28 (43)**

* Data unknown for 6 users

Table 3. Frequency of comorbidity, features of muscular dystrophy and complications of disability in 65 users with a muscular dystrophy.

Comorbidity	**Feature of MD**	**Complication of disability**
Back pain (5)	Recurrent chest infections (2)	Scoliosis (20)
Hypertension (2)	Weight loss	Dependent oedema/cellulitis (3)
Asthma (2)	Cardiomyopathy (5)	Pressure sores (2)
Cerebrovascular events (2)	Problematic pain (10)	Hip problems
Epilepsy (2)	Ventilatory failure (4)	Pelvic obliquity
Arthrogryposis	Learning difficulties	Contractures
Piles	Swallowing difficulties	
Fractures	Ureteric reflux with recurrent urinary tract infections	
Neck pain	Sleep disturbance	
Ischaemic heart disease		
Total = 18	**Total = 26**	**Total = 28**