Exploring the acceptability of implantable defibrillators in patients with cardiac dystrophinopathy and carers

Ursula M Hiermeier,1 Christine Baker,1 John P Bourke 2,3

ABSTRACT

Objective Unlike for patients with other forms of cardiomyopathies, those with severe ventricular dysfunction due to Duchenne muscular dystrophy (DMD) are not offered implantable cardioverter-defibrillator (ICD) therapy routinely. This prospective study aimed to determine the views of DMD-patients and their carers about discussing sudden death risk and their acceptance of ICDs.

Design and setting Adults with DMD (n=9) and parents/carers (n=9) participated in audio-recorded, 60–90 min focus group sessions (patients 2; parents/carers 2) conducted through either a face-to-face session at a neutral venue or a videoconference. Sessions were facilitated by a clinical psychologist, experienced in conducting focus group research. All participants understood the rationale for the study and the nature of ICD therapy. The same predefined themes were explored with each group. Recordings were transcribed, analysed thematically by two researchers, working independently and then agreed. Differences in responses between patient and carer groups were also studied and compared. Participants all provided informed written consent and the study had ethical approval.

Results Three main themes emerged: (1) access to/quality of information provided by professionals and patient engagement with them; (2) decision-making about ICDs; (3) individuals’ own ‘lived experience’ of DMD.

Conclusions The main findings were: (1) patients with DMD want to have their risk of sudden arrhythmic death discussed, when relevant and (2) if ICD therapy were established as beneficial, they would welcome an individualised discussion about its appropriateness for them.

INTRODUCTION

Duchenne muscular dystrophy (DMD) is a genetically determined, X linked recessively inherited neuromuscular disorder caused by a deficiency of the protein dystrophin on the inner aspect of cell sarcolemma. It is its clinical course is characterised by progressive weakness of proximal limb-girdle muscles and calf muscle hypertrophy. Duchenne-affected individuals lose ambulation and become wheelchair dependent before the age of 13 and—even with optimum multidisciplinary management—typically die from cardiorespiratory failure between ages 25 and 30 years. A progressive cardiomyopathy occurs in most patients with DMD. Even allowing for the improved survival achieved through better coordinated, multidisciplinary care, steroid therapy for muscle strengthening and nocturnal ventilation support, maintaining cardiac function is a prerequisite for prolonged survival.

Hearts with systolic dysfunction from any cause tend to become electrically unstable—manifesting as collapse and sudden death due to ventricular tachycardia, fibrillation or electromechanical dissociation. Implantable cardioverter-defibrillators (ICD) have been...
shown to prevent sudden arrhythmic deaths in patients with idiopathic forms of cardiomyopathy and so improve survival by detecting and treating serious heart rhythm disturbances automatically. Adults in New York Heart Association functional class II/III, with left ventricular ejection fraction below 35% due to commoner aetiologies, have long had a class I indication for ICD deployment. Not all patients benefit from device therapy, however, because of device-related complications and the impact of shock therapy.

Because DMD is a progressive disorder with widespread implications for mobility, breathing and life expectancy as well as its heart effects, the extent to which cardiac arrhythmias contribute to premature death in DMD has received little study. Furthermore, because patients remain largely free of cardiac symptoms, despite the severity of cardiomyopathy, it cannot be assumed that even discussing the possibility of sudden cardiac death with them will always be beneficial. Nor has it been established previously that they would want to have an ICD implanted, even if they were considered to be at high risk of sudden arrhythmic death. Currently, ICDs are not usually recommended to patients with DMD—primarily because of the progressive, multisystem nature of DMD, but also because it is unknown whether and to what extent devices would prolong survival meaningfully. In recent years, patients attending a dedicated cardiology-muscle clinic with advanced cardiomyopathy have had their arrhythmia risk discussed more routinely. Anecdotally, patient reactions have varied—ranging from welcoming a comprehensive discussion, including consideration of ICD therapy, to promptly curtailing the conversation. However, of the small number who have had ICDs implanted, some have received therapy appropriately for ventricular tachycardia/fibrillation, which would probably have been fatal otherwise.

The aim of this study was to determine the views of DMD-patients and parents/carers more systematically on the appropriateness of discussing arrhythmia risk and sudden death and exploring the acceptability of ICD therapy to them, if it were shown to be of benefit.

Primary outcome
The primary outcome of the research was: (1) to elicit the views of adult patients with DMD and their carers on whether they would want the risk of experiencing ventricular arrhythmias and the possibility of sudden death raised with them in discussion by their care team, when appropriate to their stage of cardiomyopathy and (2) how acceptable having a cardioverter-defibrillator implanted would be, if it were proven to prolong life in the wider DMD context.

METHODS
Participants and recruitment
The research was conducted through focus group sessions—conducted separately for DMD-patients and carers in July to August 2017. Patients aged 18 years or older, with a confirmed diagnosis of DMD, were eligible to take part in the DMD-adult group(s) and parent(s) or other adult carers were sought for the Parent group(s). Patients were not selected on the basis of known heart dysfunction and their cardiac status was not known to the research team. Participants were recruited with the help of Action Duchenne—a patient/carer support charity, through their ‘DMD-Pathfinder’ leaders, database and website. The number of participants was dictated by best practice recommendations in the conduct of thematic research studies. Once potential participants had been identified, each was invited to attend a focus group session and all received documents explaining the study and its aims. The research was made possible with the help of Newcastle upon Tyne NHS Foundation Trust charitable funds. Participants were not remunerated or reimbursed for taking part. To facilitate attendees, sessions were held on two separate occasions for each of the two groups (ie, four sessions in all). A DMD-adult and parent/carer session took place separately on each day. A face-to-face format was used for the first set and a videoconference for the second set of sessions. This change in format was to facilitate those with limited mobility, to reduce the risk of selection bias and to avoid excessive travel distance/time commitments.

Intervention and conduct of focus groups
Participants received information documents ahead of the sessions and provided written consent to taking part. Sessions began with a 30 min talk about heart involvement in DMD, the link between pumping weakness and arrhythmias and the nature and purpose of ICDs in other contexts. This was to ensure that participants were adequately briefed for the focus group session which followed. Conscious that some participants might find the topics anxiety provoking, a modified distress protocol was adopted for this research.

Analyses
Audio recordings of all focus group sessions (two DMD-adult and 2 Parent) were transcribed afterwards. Participants had the opportunity to read the transcripts of their session and correct, clarify or elaborate on their own original contribution(s). Transcripts were then analysed using an inductive thematic approach. First, two researchers studied the transcripts and labelled meaningful sections individually. Units of text with the same argument were organised and provisionally labelled into analytical categories. Results were compared between assessors and areas of disagreement resolved by consensus. Findings were organised into a hierarchy of themes and commonality and differences in responses between both DMD-adults and separately both Parent groups studied. Differences in responses between DMD-adult and Parent groups were also sought.

RESULTS
Nine adults with DMD—aged 19–48 years—took part in the DMD-adult group and, separately, nine parents (seven
Table 1A Summary of themes identified during focus group sessions

| Themes across DMD-adults and Parents | Themes captured in DMD-adults only | Themes captured in Parents only |
|--------------------------------------|------------------------------------|---------------------------------|
| **Informational care**               |                                    |                                 |
| I receive information about cardiac care from… |                                    |                                 |
| ► Routine or emergency hospital visits. | ► Parents (age related). | ► Parent support groups and networks. |
| ► Own research.                       | ► Peers.                           |                                 |
| ► Action Duchenne.                    |                                    |                                 |
| What I want to know…                 |                                    |                                 |
| ► Treatment options and interventions that might help. | ► Overview of information. | ► Advanced knowledge—issues around procedures, hospital stays, aftercare and care plan. |
| ► What is to be prioritised and at what time. | ► As much information as possible. |                                 |
| ► Mortality.                         | ► Effectiveness of treatment.       |                                 |
| ► Impacts on longevity.              | ► Future predictions (what to expect). |                                 |
| ► Risk.                              |                                    |                                 |
| ► Guidance from ‘experts by experience’. |                                    |                                 |
| **Engagement with healthcare professionals (HCP)** |                                    |                                 |
| I find it helpful when HCPs…         |                                    |                                 |
| ► Give an advanced warning so we can be prepared. | ► Are honest, but not heartless. | ► Actively involve my son. |
| ► Talk to us in sensitive, understandable and age-appropriate ways. | ► Are willing to have a detailed discussion. | ► Could address ICDs as part of routine and treatment option when ECG is introduced. |
| ► Routinely provide us with information; keep us informed. | ► Provide sufficient time to answer questions. | ► Addresses misconceptions and concerns. |
| I find it unhelpful when HCPs…       |                                    |                                 |
| ► Talk about costs.                  | ► Predict lifespan.                 | ► Do not communicate between themselves. |
| ► Make assumptions and judgements about me. | ► Withhold or present vague information; mislead me. |                                 |
| ► Talk about something without acting on it. | ► Talk about something without acting on it. |                                 |
| ► Do not drive the conversation—often the only way to find things out is to ask. | ► Provide reassurance. |                                 |
| I want HCPs to acknowledge that…     |                                    |                                 |
| ► We want and need as much information as we can. | ► Knowledge empowers me and informs my decision about ICD treatment. | ► It is up to my son. |
| ► We can adapt and adjust to what is thrown at us. | ► Information can feed worry, and I may prefer being scare informed than scared in the dark. | ► If my son is able to decide, he has the first say—please check his view and respect his decision. |
| ► We are all unique/our sons are all unique. | ► There is variation in our longevity. |                                 |
| ► We perceive our quality of life different from others’ perception of us/my son is the expert in his quality of life. | ► DMD does not stop quality of life. |                                 |
| ► There is variation from one doctor/area to another. |                                    |                                 |

female) in the Parent group. The inductive thematic analysis yielded 131 initial codes from the DMD-adult sessions, subsequently grouped on the basis of systematic review under 42 themes. Similarly, 102 codes were identified from the Parent group and organised under 31 themes. Thirty-nine themes were common to both groups (Table 1A, B). The themes identified were categorised under three main headings: (1A) access to and quality of information provided by medical professionals and (1B) patient engagement with healthcare professionals (HCP); (2) decision-making about ICDs; (3) individuals’ own lived experience with DMD.

Informational care and the engagement with HCPs
Parents and DMD-adults demonstrated only limited understanding of ICDs and their possible role in the management of advanced cardiomyopathy. In discussion, both DMD-adults and Parents raised many questions (box 1).

Patients and parents/carers share the information they derive from various sources about care requirements of
Table 1B   Themes identified during focus group sessions

| Themes across DMD-adults and Parents | Themes captured in DMD-adults only | Themes captured in Parents only |
|--------------------------------------|-----------------------------------|---------------------------------|
| Decision-making: weighing up the pros and cons about having an ICD | Benefits of having an ICD | Avoid complexity of ‘normal’ resuscitation. |
| ► Prolong life. | ► Easy to maintain for my family/carer.s. | ► Preventative measure. |
| ► Improve life expectancy. | ► There is a time when I am ready to think about this; it is not yet (age related). | ► Past experience; we have seen long-term benefits of other decisions and treatments. |
| ► Improved quality of life. | ► Avoid risk of death—not having operation is also a risk. | ► Considered to be safe. |
| ► Reassurance. | ► Avoid complexity of ‘normal’ resuscitation. | ► Another treatment option. |
| ► Avoid risk of death—not having operation is also a risk. | ► Easy to maintain for my family/carer.s. | ► My life is better when his life is better. |

Concerns about having an ICD

| ► Risk—every operation carries risk. | ► Risk of anaesthetic and operation. | ► Triggers thoughts about mortality. |
| ► Impact of being hospitalised—hospital staff do not understand needs, and carers are not able to come and help. | ► Reoccurring battery change. | ► My son may be affected by information. |
| ► Sudden death. | ► Right age for procedure. | ► Aftercare will be demanding. |
| ► Can we go through hospital stay (again). | ► Pain. | ► This is another slap in the face. |
| ► Uncertainty. | ► Scared of ECG. | ► I am getting older. |

Things to be weighed up

| ► Bother of the procedure. | ► Perception of general health. | ► How does my son feel about ICD. |
| ► Perception of general health. | ► State of heart deteriorated. | ► I accept uncertainty. |
| ► Personal choice and preference. | ► I take responsibility for my body. | ► I accept uncertainty. |

I will adjust and cope!

| ► Fear is part of life, initial anxiety passes. | ► I take responsibility for my body. | ► I accept uncertainty. |
| ► I take things as they come. | ► I get used to devices and equipment in my body. |  |
| ► I have coped with worse, I will adapt. |  |  |

Not part of the decision process

| ► Costs of an ICD. | ► My family—they will be reassured. | ► I am not spooked about having a device in my body; I trust mechanical things. |
| ► My family—they will be reassured. | ► I am not spooked about having a device in my body; I trust mechanical things. |  |
| ► Switching off the ICD (only if quality of life down or in pain). |  |  |

My own experience

| As DMD adult… | As parent… |
|---------------|------------|
| ► I acknowledge my mortality. | ► I want to protect my son. |
| ► I want to live as long as possible. | ► I want to avoid overburdening my son, for example, with information. |
| ► I need the right treatment at the right time to extend my life. | ► I do not like to leave my son for too long. |
| ► I want to live to my potential and promote my quality of life. | ► I need to be on top of my son’s care. |
| ► I prefer a sudden shock to a sudden death. | ► I have a different experience—I am nurse, physiotherapist, line manager, pharmacist, carer and parent, while my own life needs relationships, privacy, home time, jobs, holidays and flexibility. |
| ► Risk of dying is worse than having a bad quality of life. | ► Your life is ‘on hold’—I live day by day. |
|  | ► I have a certain ‘timeline expectation’. |

DMD, Duchenne muscular dystrophy; ICD, implantable cardioverter-defibrillator.

DMD (table 1A, B). However, the most valuable source of information from their perspectives would be that provided by experts and by patients who had already received ICDs and their families. All groups felt it important that, when clinical information was provided, it was communicated in sensitive, understandable and age-appropriate ways—both in overview and in depth.

Some Parents expressed concerns that information might increase their sons’ anxiety, while others felt that their son was open to discussion on all topics. DMD-adults considered it better to be aware of possibilities of change in their condition than to worry about uncertainties inherent to it (quote 1—refer to online supplementary appendix). None could identify any circumstances in
Box 1 Questions raised by DMD-adults and Parents during focus group sessions

1. How do you know when the heart is not in rhythm?
2. Is it better having them at a certain age?
3. How does an ICD work? How does it control your heart?
4. How does the ICD work in terms of recording and tracking heart rhythm and sending this to the hospital?
5. How many DMD adults have an ICD? How is it working for them?
6. How big is the device?
7. Can you experience how it feels to get a shock before you have an ICD implanted?
8. Can you put the ICD in at the same time as spinal surgery?
9. How does the operation look like? What does it entail?
10. When does the ICD ‘jump’ in?
11. How does an ICD feel when it is ‘in action’?
12. How many times will the ICD go off?
13. Would you ring an ambulance straight away?
14. Will the alarm go off when you go through the metal detectors at the airport with having an ICD?
15. How easy is it to change the battery?
16. How would you turn the ICD off? Can you turn it back on?
17. If you want an ICD, how would you go about getting one? Can an ICD be part of a routine cardiac protocol?
18. How many times will the ICD go off?

DMD, Duchenne muscular dystrophy; ICD, implantable cardioverter-defibrillator.

which they would not want to be informed about the state of their heart since they viewed knowledge as empowering, allowing them to feel in control and allowing them to be proactive in making decisions about their management.

DMD-adults were of the view that, if they wanted information from HCPs, they had to ask for it specifically and ‘drive’ the conversation to get answers. They also felt that often there was not enough time to raise important questions at specialist review appointments.

Participants in all groups emphasised their desire for open and honest communication—in particular about life and death issues. All expressed an existential need for more information about their/sons’ longevity and quality of life.

DMD-adult: …To me it is important to know all the facts about all treatments, because you need to know what to do to try and live longer. Because you need to have the right treatment at the right time, because it can be poorer treatment if you get it wrong…. Accessing appropriate treatments at the right time was seen as vital by all participants and considered critical to achieving prolonged survival by both DMD-adult groups. However, given the uncertainties, DMD-adults did not want to hear predictions about their lifespan.

DMD-adult: …In terms of what you don’t want to know—you don’t want a ‘sell-by’ date given. The whole idea ‘you will not survive past that date’, because I find that more depressing than anything else…

While acknowledging that not all patients required or wanted the same amount of information about their management, DMD-adults expected HCPs to acknowledge the uniqueness of each individual and the particular circumstances of each family in their discussions—rather than making unhelpful generic assumptions about them. Both participant groups felt that HCPs did not communicate effectively with each other. In their experience, cardiac treatments seemed to vary with locality and specialist. Similarly, parents had concerns that some HCPs would not necessarily be aware or inform them of changes or innovations in treatments for their condition—such as about ICDs (quote 4—see online supplementary appendix). All groups discussed variations in the standard of NHS care provided between different parts of the country and, what they saw as, a lack of uniformity in the cardiac treatment of DMD.

Decision-making: pros and cons of ICDs and ‘Lived experiences’

Both DMD-adult and Parent groups were open to the idea of new technology. All viewed the concept of ICD therapy positively, feeling that device implantation would provide reassurance, protection and empowerment (quote 5—see online supplementary appendix). DMD-adults were interested in their sons having ICDs implanted. As long as their son was competent and old enough, Parents expressed support for their sons’ decisions and wanted them to have the final say.

All four group discussions accepted that there were risks inherent in most health-related decisions and that operative risk was hard to quantify:

DMD-adult: …I think for me, it is not straight forward, because obviously you got the risk of the procedure, but you also got the risk of not having the procedure, and then, you got the risk of what happens if you get to a certain point when you can’t actually have the surgery, and that is another risk….

Both parents and parents viewed the decision about having ICD treatment to be a balance between quality of life and prolonging life (quote 7—see online supplementary appendix). However, concerns about the implant procedure were raised because of previous bad experiences following general anaesthesia and they had similar concerns about repeat procedures for generator replacement (quote 8—see online supplementary appendix). Parents expressed fears about the degree of postoperative pain associated with the implant procedure. Further issues were raised around critical shortcomings in the care received from ward staff during hospital admissions when their own carers were not allowed to be with them.

The impact of an ICD on the lives of patients with DMD was discussed. Some expressed the view that anything was preferable to sudden death and so did not consider that fear of shock therapy or of the pain they might experience from a shock would affect their decision to have the device. The consensus was that they would adjust to an
ICD—just as they had done for other invasive treatments and equipment previously.

**DMD-adults** …Yes, I think, quality of life is quite subjective. You can adapt quite well to whatever is thrown at you. In a way, I just want to prolong live as long as I can and then work out afterwards what my quality of life is…

Parents held similar views about their own ability to adjust to any adversity DMD might bring (quote 10—see online supplementary appendix). Nor did **DMD-adults** consider it likely that having an ICD would change their sense of who they were. **Parents** were worried that their sons might become ICD obsessed or paranoid because of the implanted device, wondered whether day-to-day anxieties might be increased and how well their sons would react to receipt of shock therapy. Overall, **DMD-adults** thought it likely that their family would react positively despite initial unease. They concluded that ICDs would reassure parents/carers and give them peace of mind.

Some parents felt that their son’s quality of life was already limited, citing examples like being unable to drive a car, having partners or meeting other people, so **DMD-patients** felt that outsiders often underestimated their quality of life, which they felt was generally good. Several **Parents** admitted to being continuously fearful about their sons dying:

**Parent** …I think, I am in denial. You know, I am blocking it all. So, yes, I am like that, I am scared. I don’t want him to go… I am scared…

**Parent** …No, no, no… you don’t know what happens in two weeks. I am in denial. I think that is a form of protection for me. I am quite happy like that to be fair…

**Parent** …I think because, I don’t know, over the years of seeing how things can just change so quickly. He can become ill, he’d get seriously ill very quickly, you sort of just live for the day … you don’t plan years ahead … You just carry on the way you do. Get on with life every day and not sort of focus too much about everything that is going on. You just have to enjoy the time that you are together and…

**Parent** …you are right.…

**Parent** …yes, and then you go to [a specialist] and they say, ‘your boy is living until they are 50.’ And you are like, ‘shit… that is really good news, but… shit, because I’ll be 80…’.

**DMD-adults** were also asked under what circumstances they might decide to have their ICD deactivated (ie, have therapies turned off, without need to have the ICD explanted). Some questioned the need for this decision altogether, since the rationale for an ICD is to prolong life; others would consider deactivation if life became more painful if they experienced serious device or DMD-related complications (quote 12—see online supplementary appendix). **Parents** voiced concerns about the burden that an ICD deactivation decision would have on their sons. In discussing the cost of ICD therapy and whether this affected decisions to implant them, both adults and parents concluded that cost was not an important determinant of this therapy (quote 13—see online supplementary appendix).

**DISCUSSION**

The aim of this research was not to establish the utility of ICD therapy in patients with DMD but to explore patient and carer views of the benefits or otherwise of discussing arrhythmia risk and whether they would want to be considered for prophylactic device therapy, if it were shown to be beneficial.

Worldwide, a small number of patients with cardiac dystrophinopathy have had ICDs implanted for prophylactic indications and there are anecdotal reports of some receiving antitachycardia pacing or shock therapy appropriately for spontaneous arrhythmias.24 However, whether widespread deployment of ICDs would prolong life for the majority of patients with DMD remains to be determined.25 It is also uncertain whether the increased risk of the implant procedure itself justifies the potential benefits in this progressive, multisystem condition.26 27 From the limited information available, arrhythmia risk seems low during most of the course of cardiomyopathy in DMD and sustained ventricular arrhythmias seem only to occur with the onset of overt heart failure symptoms.15–19

As summarised, the four focus groups provided a wealth of themes and an overall positive response from both **DMD-adult** and **Parents** groups to discussing arrhythmia risk and the concept of ICD therapy. Patient groups voiced a general concern that medical personnel, family and friends often underestimated and misjudged their quality of life—which they felt was good.28 29 Although anticipating deteriorating health, they valued life-extending therapies in general and so were hopeful about the potential benefits of ICDs in that regard. However, they had many questions and concerns about the procedural risks of the implant procedure itself and how receiving shock therapy from an ICD might affect them. Parent/carer groups shared fears about surgery in general, the need for any hospital stay and unmet post-procedure recovery needs of their sons. They felt that ICDs could provide reassurance in the long run and felt hopeful that it might improve the health and longevity of their sons’ lives. They also felt it would put their own minds more at ease.

Both **DMD-adult** and **Parents** groups stressed the importance of having appropriate, timely information to aid them in all their clinical decision-making. Some of the strongest views expressed were about deficiencies in the way information was provided. It was often felt to be conveyed in a generic, hurried and insensitive manner—without allowing adequate time for discussion and not tailored to individual patient differences or context. When conveyed appropriately, patients considered information to be empowering—giving them a greater feeling...
of being in control and allowing them to be proactive in decision-making.

The reaction of DMD-adults to the focus group sessions was positive. They felt their sessions were interesting and thought provoking. Indeed, some stated that they were happy to be ‘an experiment generation’ and, if appropriate, to trial ICDs, for the benefit of future generations. Although discussing these topics seemed at times emotional and complex, participants agreed that it was really helpful and enjoyable.

Limitations

By its nature, thematic research can only legitimately explore the views of a small sample of eligible participants, whose views need not necessarily represent those of the wider groups from which they are derived. In addition, adult patients with DMD have limited mobility and are reliant on carers for activities such as, for example, travelling to a face-to-face group session. It was in recognition of this potential participant selection bias that one of the focus group sessions was conducted by videoconference. The consistency in themes identified in the face-to-face and teleconference format adult patient group sessions provides further reassurance that the groups did not differ significantly in participant views and opinions.

CONCLUSIONS

The two main conclusions from this research were, first, that adult patients with DMD wanted to have the possibility of sudden cardiac death discussed when relevant to their stage of cardiomypathy and, second, if ICD therapy were established as beneficial, they would welcome a detailed discussion about its risks and benefits—individualised to them. Since ICD deployment affects end-of-life decisions, the discussion should involve patients, caregivers and family and follow a shared decision-making model. Cardiologists providing care for patients with advanced cardiac dysfunction should take account of these novel findings, derived from patient and carer perspectives.

Acknowledgements

The authors gratefully acknowledge the help of Action Duchenne, UK, in encouraging and facilitating this research and the DMD-Pathfinder group in assisting with recruitment.

Contributors

UMH: study design, recording and transcribing focus group sessions, thematic analysis, manuscript preparation, revision for content and proofing. CB: study design, leading and facilitating focus group sessions, thematic analysis, manuscript preparation, revision for content and proofing; responsible for the overall content as guarantor of all thematic analyses. JPB: study concept, protocol design and data acquisition, obtaining funding, study supervision, interpretation of data, drafting manuscript for content, final approval of manuscript, accountable for all aspects of the research.

Funding

The research team received funding support from ‘Charitable Funds’ at Newcastle upon Tyne Hospitals NHS Foundation Trust. Study sponsor: Newcastle upon Tyne Hospitals NHS Foundation Trust, UK, was the NHS sponsor for this research.

Disclaimer

Charitable Trustees did not have any role in study design, data analysis, results interpretation or manuscript preparation. The Trust has not had any role in the design of the study or its protocol, nor did it play any part in the analysis of results or had influence/authority over the presentation of its findings.

Competing interests

JPB reports funding support from ‘Charitable Funds’, NUTH NHS Foundation Trust for this research and a grant from British Heart Foundation for another DMD-related research study ongoing over the same time period.

Patient consent for publication

Not required.

Ethics approval

Ethics approval for the study was granted by ORECI-Northern Ireland Ireland Committee, UK (IRAS project ID: 215892).

Provenance and peer review

Not commissioned; externally peer reviewed.

Data availability statement

All data relevant to the study are included in the article or uploaded as supplementary information. Additional participant quotes on which the analysis has been based are either included in the manuscript or in the appendix supplied as part of this submission.

Open access

This is an open access article distributed in accordance with the Creative Commons Attribution Non Commercial (CC BY-NC 4.0) license, which permits others to distribute, remix, adapt, build upon this work non-commercially, and license their derivative works on different terms, provided the original work is properly cited, appropriate credit is given, any changes made indicated, and the use is non-commercial. See: http://creativecommons.org/licenses/by-nc/4.0/

ORCID iD

John P Bourke http://orcid.org/0000-0001-7857-9073

REFERENCES

1 Birnkranjt DJ, Bushby K, Barrn CM, et al. Diagnosiss and management of Duchenne muscular dystrophy: part 1: diagnosis, and neuromuscular, rehabilitation, endocrine, and gastrointestinal and nutritional management. Lancet Neurol 2018;17:251–67.

2 Arora H, Willockocks R, Lott DJ, et al. Longitudinal timed function tests in Duchenne muscular dystrophy: Imaging DMD cohort natural history. Muscle Nerve 2016;58:631–8.

3 Passamano L, Taglia A, Palladino A, et al. Improvement of survival in Duchenne muscular dystrophy: retrospective analysis of 835 patients. Acta Myol 2012;31:121–5.

4 Moxley RT, Pandya S, Ciatofoni E, et al. Change in natural history of Duchenne muscular dystrophy with long-term corticosteroid treatment: implications for management. J Child Neurol 2010;25:1116–29.

5 Kamdar F, Garry DJ. Cardiomyopathy D-D. Dystrophin-Deficient cardiomyopathy. J Am Coll Cardiol 2016;67:2533–46.

6 McNally EM, Kaltman JR, Benson DW, et al. Contemporary cardiac issues in Duchenne muscular dystrophy. Working group of the National heart, lung, and blood Institute in collaboration with parent project muscular dystrophy. Circulation 2015;131:1590–8.

7 Saito T, Kawai M, Kimura E, et al. Study of Duchenne muscular dystrophy long-term survivors aged 40 years and older living in specialized institutions in Japan. Neuromuscul Disord 2017;27:107–14.

8 Kauppila JP, Hantula A, Kortelaainen M-L, et al. Association of initial recorded rhythm and underlying cardiac disease in sudden cardiac arrest. Resuscitation 2018;125:76–8.

9 Suryanarayana P, Garza H-HK, Klewer J, et al. Electrophysiological considerations after sudden cardiac arrest. Curr Cardiol Rev 2018;14:102–8.

10 Moss AJ, MADIT-I and MADIT-II. J Cardiovasc Electrophysiol 2003;14:596–8.

11 Bardy GH, Lee KL, Mark DB, et al. Amiodarone or an implantable cardioverter-defibrillator for congestive heart failure. N Engl J Med 2005;352:225–37. [SCD-HeFT].

12 El Mohed M, Nicolas J, Khannis AM, et al. Implantable cardiac defibrillators for people with non-ischaemic cardiomyopathy. Cochrane Database Syst Rev 2018;12:CD012738.

13 Kusumoto FM, Bailey KR, Chauki AS, et al. Systematic review for the 2017 AHA/ACC/HRS guideline for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. Heart Rhythm 2018;15:e253–74.

14 Olde Nordkamp LRA, Postema PG, Knops RE, et al. Implantable cardioverter-defibrillator harm in young patients with inherited arrhythmia syndromes: a systematic review and meta-analysis of inappropriate shocks and complications. Heart Rhythm 2016;13:443–54.

15 Yotsukura M, Ishizuka T, Shimada T, et al. Late potentials in progressive muscular dystrophy of the Duchenne type. Am Heart J 1991;121(4 pt 1):1137–42.
16 Himmrich E, Popov S, Liebrich A, et al. [Hidden intracardiac conduction disturbances and their spontaneous course in patients with progressive muscular dystrophy] [German]. Z Kardiol 2000;89:592–8.

17 Menon SC, Etheridge SP, Liesemer KN, et al. Predictive value of myocardial delayed enhancement in Duchenne muscular dystrophy. Pediatr Cardiol 2014;35:1279–85.

18 Villa CR, Czosek RJ, Ahmed H, et al. Ambulatory monitoring and arrhythmic outcomes in pediatric and adolescent patients with Duchenne muscular dystrophy. J Am Heart Assoc 2015;5:e002620.

19 Chiang DY, Allen HD, Kim JJ, et al. Relation of cardiac dysfunction to rhythm abnormalities in BMD. Am J Cardiol 2016;117:1349–54.

20 Canter CE, Shaddy RE, Bernstein D, et al. Indications for heart transplantation in pediatric heart disease: a scientific statement from the American heart association Council on cardiovascular disease in the young; the councils on clinical cardiology, cardiovascular nursing, and cardiovascular surgery and anesthesia; and the quality of care and outcomes research interdisciplinary Working group. Circulation 2007;115:658–76.

21 Vaismoradi M, Turunen H, Bondas T. Content analysis and thematic analysis: implications for conducting a qualitative descriptive study. Nurs Health Sci 2013;15:398–405.

22 Castileberry A, Nolen A. Thematic analysis of qualitative research data: is it as easy as it sounds? Curr Pharm Teach Learn 2018:10:807–15.

23 Draucker CB, Martosof DS, Poole C. Developing distress protocols for research on sensitive topics. Arch Psychiatr Nurs 2009;23:343–50.

24 Fragakis N, Sotiriadou M, Krexi L, et al. Electrical storm in a patient with Duchenne muscular dystrophy cardiomyopathy triggered by abrupt β-blocker interruption. Ann Noninvasive Electrocardiol 2017;22. doi:10.1111/ane.12477. [Epub ahead of print: 15 May 2017].

25 Buddhe S, Cripe L, Friedland-Little J, et al. Cardiac management of the patient with Duchenne muscular dystrophy. Pediatrics 2018;142:S72–81.

26 Fayssoil A, Lazarus A, Wahbi K, et al. Cardiac implantable electronic devices in tracheotomized muscular dystrophy patients: safety and risks. Int J Cardiol 2016;222:975–7.

27 Shakya S, Matsui H, Fushimi K, et al. In-Hospital complications after implantation of cardiac implantable electronic devices: analysis of a national inpatient database in Japan. J Cardiovasc Comput Tomogr 2017;10:405–10.

28 Rahbek J, Werge B, Madsen A, et al. Adult life with Duchenne muscular dystrophy: observations among an emerging and unforeseen patient population. Pediatr Rehabil 2005;8:17–28.

29 Albrecht GL, Devlieger PJ. The disability paradox: high quality of life against all odds. Soc Sci Med 1999;49:977–88.

30 Lewis KB, Carroll SL, Birnie D, et al. Incorporating patients’ preference diagnosis in implantable cardioverter defibrillator decision-making: a review of recent literature. Curr Opin Cardiol 2018;33:42–9.