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

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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.

Key questions

What is already known about this subject?

► A progressive cardiomyopathy affects almost all patients with Duchenne muscular dystrophy (DMD). Unlike in patients with severe ventricular dysfunction of other aetiologies, arrhythmia risk and prophylactic implantable cardioverter-defibrillator (ICD) therapy are not discussed routinely in DMD because the benefits are unknown and there are concerns about causing needless distress.

What does this study add?

► Contrary to some healthcare professionals' assumptions, patients want to have individualised discussions both about the possibility of sudden cardiac death and the risks/benefits of prophylactic ICD implantation, when relevant to their stage of cardiomyopathy.

How might this impact on clinical practice?

► Patients with DMD and their families want healthcare professionals to be more open in communications with them generally. Some expressed willingness to be 'an experimental generation' and to trial ICDs for the benefit of clarifying their utility in DMD.

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.[1 2] 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.[3 4] A progressive cardiomyopathy occurs in most patients with DMD.[5 6] 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.[7]

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.[8 9] 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.[10–12] 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 1 indication for ICD deployment.[10 13] Not all patients benefit from device therapy, however, because of device-related complications and the impact of shock therapy.[14]

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.[15–19] 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.[20] 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.[20] 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.[21 22] 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.[23]

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.[21 22] 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. ► Own research. ► Action Duchenne.	► Parents (age related). ► Peers.	► Parent support groups and networks.
What I want to know…		
► Treatment options and interventions that might help. ► What is to be prioritised and at what time. ► Mortality. ► Impacts on longevity. ► Risk. ► Guidance from 'experts by experience'.	► Overview of information. ► As much information as possible. ► Effectiveness of treatment. ► Future predictions (what to expect).	► Advanced knowledge—issues around procedures, hospital stays, aftercare and care plan.
Engagement with healthcare professionals (HCP)		
I find it helpful when HCPs…		
► Give an advanced warning so we can be prepared. ► Talk to us in sensitive, understandable and age-appropriate ways. ► Routinely provide us with information; keep us informed.	► Are honest, but not heartless. ► Are willing to have a detailed discussion. ► Provide sufficient time to answer questions. ► Are specialists.	► Actively involve my son. ► Could address ICDs as part of routine and treatment option when ECG is introduced. ► Addresses misconceptions and concerns. ► Have separate, first conversation with parents (age dependant).
I find it unhelpful when HCPs…		
► Talk about costs.	► Predict lifespan. ► Make assumptions and judgements about me. ► Withhold or present vague information; mislead me. ► 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.	► Do not communicate between themselves.
I want HCPs to acknowledge that…		
► We want and need as much information as we can. ► We can adapt and adjust to what is thrown at us. ► We are all unique//our sons are all unique. ► We perceive our quality of life different from others' perception of us//my son is the expert in his quality of life. ► There is variation from one doctor/area to another.	► Knowledge empowers me and informs my decision about ICD treatment. ► Information can feed worry, and I may prefer being scare informed than scared in the dark. ► There is variation in our longevity. ► DMD does not stop quality of life.	► It is up to my son. ► If my son is able to decide, he has the first say—please check his view and respect his decision.

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		
► Prolong life. ► Improve life expectancy. ► Improved quality of life. ► Reassurance. ► Avoid risk of death—*not* having operation is also a risk.	► Easy to maintain for my family/carers. ► There is a time when I am ready to think about this; it is not yet (age related).	► Avoid complexity of 'normal' resuscitation. ► Preventative measure. ► Past experience; we have seen long-term benefits of other decisions and treatments. ► Considered to be safe. ► Another treatment option. ► *My* life is better when his life is better.
Concerns about having an ICD		
► Risk—every operation carries risk. ► Impact of being hospitalised—hospital staff do not understand needs, and carers are not able to come and help. ► Sudden death. ► Can we go through hospital stay (again). ► Uncertainty.	► Risk of anaesthetic and operation. ► Reoccurring battery change. ► Right age for procedure. ► Pain. ► Scared of ECG. ► My family—they might be worried; extra burden.	► Triggers thoughts about mortality. ► My son may be affected by information. ► Aftercare will be demanding. ► This is another slap in the face. ► I am getting older. ► Anxiety might escalate cardiac deterioration. ► My son might think everyone had enough and not go on.
Things to be weighed up		
► Bother of the procedure.	► Perception of general health. ► State of heart deteriorated. ► Personal choice and preference.	► How does my son feel about ICD.
I will adjust and cope!		
► Fear is part of life, initial anxiety passes. ► I take things as they come. ► I have coped with worse, I will adapt.	► I take responsibility for my body. ► I get used to devices and equipment in my body.	► I accept uncertainty.
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. ► 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 live as long as possible. ► I need the right treatment at the right time to extend my life. ► I want to live to my potential and promote my quality of life. ► I prefer a sudden shock to a sudden death. ► Risk of dying is worse than having a bad quality of life.	► I want to protect my son. ► I want to avoid overburdening my son, for example, with information. ► I do not like to leave my son for too long. ► I need to be on top of my son's care. ► 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. ► 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 get 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 DMD adults would an ICD be right for?

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/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 patients 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-adult*: …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, but *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 of 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 cardiomyopathy 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.[30] Cardiologists providing care for patients with advanced cardiac dystrophinopathy should take account of these novel findings, derived from patient and carer perspectives.

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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.

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