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1 Review

2 Title: An integrative review exploring psycho-social impacts 3 and therapeutic interventions for parent caregivers of young 4 people living with Duchenne's Muscular Dystrophy.

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11

12 **Abstract:** The purpose of this integrative review was to explore psycho-social impacts and
13 therapeutic interventions for parent caregivers of young people living with Duchenne's Muscular
14 Dystrophy (DMD). This review utilised Whittemore and Knaf'l's [1] methodology. Electronic
15 databases were searched for research publications between 2010 and 2020. This included Medline,
16 CINAHL, PsycINFO, ERIC, ERC, AMED. Four central themes emerged: Living with DMD;
17 Knowing and telling; Transitioning; Building resilience. The impact on parents caring for a child
18 with DMD affected all aspects of their lives, changed over time and had identifiable peak stress
19 points. Unmet parental information and support needs left parents struggling in their role.
20 Transition required changes to parenting behaviours and required adaptation and resilience. It is
21 proposed that future investment should focus on anticipating family need, targeting intervention
22 cognisant of predictable stress points and building resilience through social community. Parents
23 may then be better positioned to support their child in looking forward.

24 **Keywords:** Psycho-social needs; Muscular Dystrophy; **Duchenne's Muscular Dystrophy**; Parents
25 Caregivers; Age 13-19; Family burden/Care burden
26

1. Introduction

Duchenne Muscular Dystrophy (DMD) is an X-linked, degenerative, neuromuscular disorder with an estimated male birth incidence of 1:3800 to 1:6200 [2]. The disease is characterised by a progressive degeneration of muscle fibres resulting in muscle weakness and eventual loss of ambulation. Functional dependence typically occurs in the second decade of life [3] with cardiac and respiratory complications often shortening life. Other types of muscular dystrophies, such as, Becker's muscular dystrophy (BMD) and limb-girdle muscular dystrophy (LGMD) have similar progression to DMD but a near normal life expectancy with symptoms appearing later, being less severe and thus preserving ambulation often to mid-life.

DMD as the most common and severe form of the disease is the focus for this review. Disruption to daily life with DMD can commence at an early age and not only impacts the child but also the family. The nature of the disease itself and care required places a heavy burden on parent caregivers for an extended period of their lives. Improvements in supportive care for children and young people with DMD have resulted in improved quality of life and life expectancy but until recently drug therapies had seen little change. This situation is rapidly changing with the emergence of new therapies that address underlying genetic defects setting a change in course for DMD treatment [2]. Antisense oligonucleotides (ASO), are new therapies that modify disease pathways by targeting underlying genetic changes. There is now potential to prevent clinical features of disease occurring with early intervention. [3], [4]. Another new treatment is Ataluren (Translarna), this is not an ASO but this drug demonstrates that the non-sense mutation a genetic defect causing some types of DMD, can respond to treatment [4]. In a disease that has been seen as incurable for so long, these novel treatments offer a much-improved outlook for the future.

The aim of this integrative review is to explore psycho-social needs and therapeutic interventions for parent caregivers of young people living with muscular dystrophy.

2. Purpose and Questions

2.1. Purpose of the study

This integrative review addressed the psycho-social needs and therapeutic interventions for caregivers of young people living with muscular dystrophy.

2.2. Research questions

(1) What is the psycho-social impact on caregivers of young people living with muscular dystrophy?

(2) What therapeutic interventions are employed to meet the psycho-social needs of caregivers of young people living with muscular dystrophy?

(3) What factors influence accessibility of psycho-social interventions for these caregivers?

3. Method

The integrative review method systematically summarises and analyses empirical and theoretical literature to provide an in-depth understanding of a problem or phenomenon for the purpose of providing an evidence base to practice and policy development [7]. Developed to enhance rigour in the conduct of any review, Whittemore and Knaf's [1] modified five stage integrative review framework can provide a systematic and rigorous structure to conduct an integrative review [7,8]. This review adopted this framework and consistent with this followed the framework stages: 1. Problem identification, 2. Literature search, 3. Data evaluation, 4. Data analysis, and 5. Presentation of results [1].

3.1. Literature search

Integrative reviews use more than one search strategy to enhance the quality of the review and minimize incomplete and biased results [1,7,8]. Computerised databases may only yield approximately 50% eligible studies [8]. Within this study two search strategies were made. The search was undertaken in November 2020 using a combination of the following key words: Psych-social needs, Muscular Dystrophy, **Duchenne Muscular Dystrophy**, Parent Caregivers, Mothers, Fathers, Family, Psycho-social needs / Interventions, Age 13-19, Family burden/Care burden. **Use of Muscular Dystrophy as a key term enabled a broad search ensuring capture of all relevant papers prior to a selective search using Duchenne Muscular Dystrophy.**

Search strategy 1: Electronic databases were searched for research publications between 2010 and 2020. This included Medline, CINAHL plus, PsycINFO, ERIC, ERC, AMED. Individual databases: British Nursing Index, DARE, Cochrane Library, Joanna Briggs Institute, EThOS also completed. A manual search was conducted of the reference lists of the identified articles. To focus on the most recent publications and to identify any new emerging data the search was limited to material available between 2010-2020. Inclusion criteria identified data that addressed the aims of the study which included psycho-social needs and factors that influence accessibility of psycho-social interventions. This was achieved initially by reviewing the titles and abstracts. Any data collected that did not meet the eligibility criteria was excluded **Table 1**. Studies that met the inclusion criteria were reviewed and organised into a table.

Search strategy 2: Hand searching reference lists of retrieved articles to find relevant literature not previously identified, including grey literature. **Grey literature is documentation types that includes government, academics, business, and industry** in print and electronic formats that are protected by intellectual property rights.

Examples of grey literature include conference abstracts, presentations, proceedings; regulatory data; unpublished trial data; government publications; reports (such as white papers, working papers, internal documentation); dissertations/theses; patents and policies [9,10]. All data accessed was excluded from the search as in the main they were secondary sources of data or websites with resources to access.

Table 1. Inclusion and exclusion criteria.

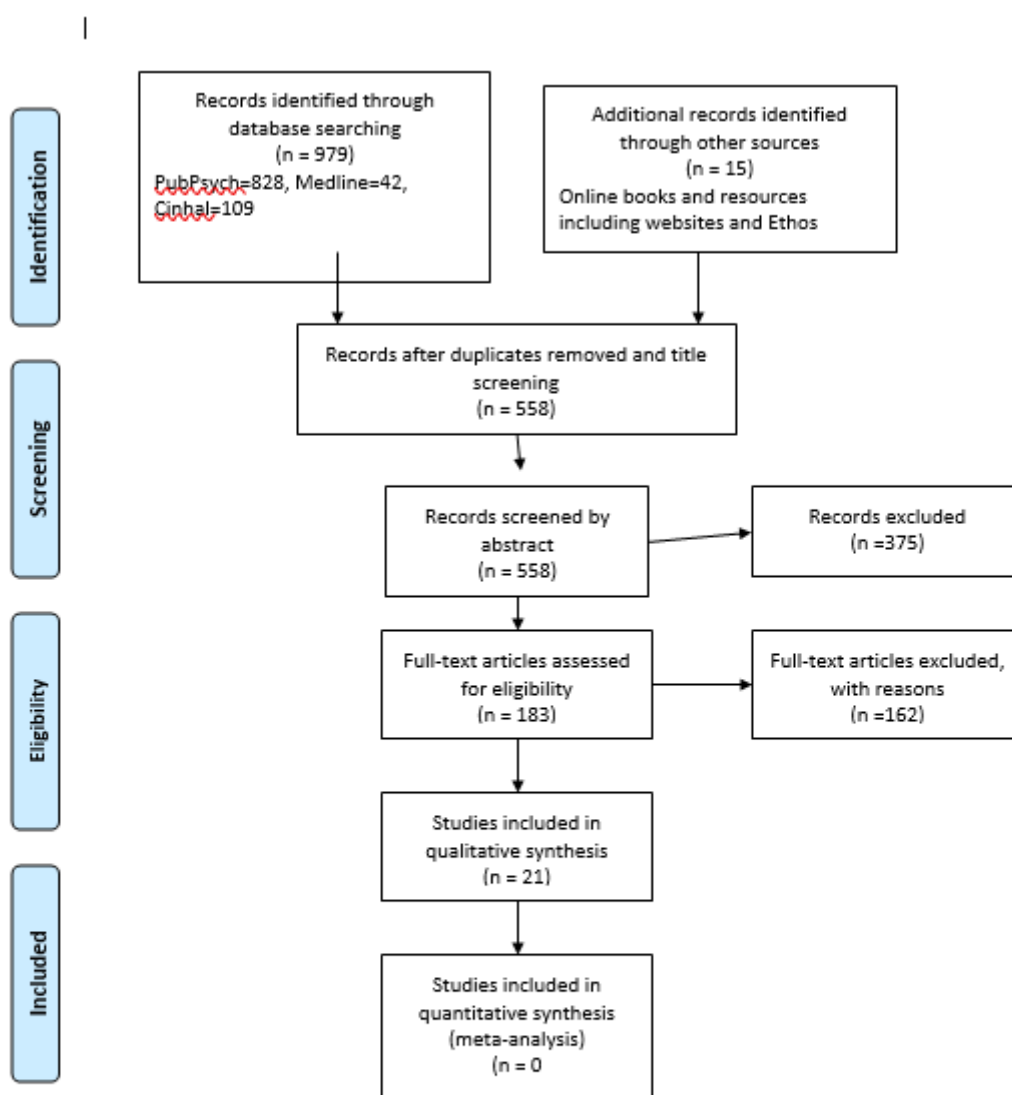
Inclusion criteria	Exclusion criteria
Primary sources 2010-2020	Paper in press
Psycho-social needs	
Factors that influence accessibility of psycho-social interventions	Abstracts, conference proceedings
Original publication in English	Websites with resources available
Meets aims of study	Secondary source or meta literature

3.2. Description of the studies

The search strategy identified 979 articles: PubPsych 828, CINAHL 409, Medline 42. Additional resources identified as grey literature, 15 including government policies and Ethos. Following screening of the titles and removing duplicates (558), articles remained. Abstracts of the remaining articles were reviewed and if they did not adhere to the aims of the study they were excluded. The total selected for the review was 21. The literature search and the screening of the research was undertaken by the authors (D.P), (C.E) and (B.D). Verification was undertaken by (J.A). The study designs of the chosen articles were qualitative and addressed the aims of the study.

119

Table 2.



From: Moher D, Liberati A, Tetzlaff J, Altman DG, The PRISMA Group (2009). Preferred Reporting Items for Systematic Reviews and Meta-Analyses: The PRISMA Statement. *CMAJ* Med 6(7): e1000097. doi:10.1371/journal.pone.000097

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3.3. Quality Appraisal of the chosen studies

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As part of any appraisal of literature it is important to assess the methodological quality of a study. The focus is to assess the extent which a study may be bias in its design, conduct and analysis. In this review we applied Joanna Briggs Institute (JBI) quality appraisal checklist for qualitative research [8]. All studies chosen were subjected to rigorous appraisal and reviewed independently by the researchers. The results of this appraisal then were used to inform synthesis and interpretation of the results of this study.

128

3.4. Data analysis

129

130

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132

As part of the data reduction a spreadsheet was developed. The data reduction commenced by assessing each article for its relevance to the study aims. Key themes then emerged, then the following stage of analysis was to identify sub-themes. These sub-themes were then analysed and synthesized. For each individual piece of data, we

133 identified any duplications, discussed if the article met the study aims and reached
134 consensus of significant items.

135
136 **Four key themes with additional sub themes were identified. These are *Living with***
137 ***DMD, Knowing and telling, Transitioning, and Building resilience.***

138 139 **4. Main Findings**

140
141 This integrative review posed questions around impact, intervention and
142 accessibility. The studies analysed were able to provide insight into the extensive psycho-
143 social impacts on parent caregivers of young people living with muscular dystrophy.
144 Three of the themes which emerged *Living with DMD, Knowing and telling, and*
145 *Transitioning* represent the challenges experienced.

146 In contrast there was limited evidence, within the reviewed papers, relating to the
147 accessibility of the therapeutic interventions used to support families. Interventions
148 were often only considered when making suggestions for future research and
149 developments. The final theme, *Building Resilience*, reflects how the studies provided
150 some description of interventions employed but what is not addressed is accessibility.

151
152 In the majority of studies exploring parental experiences, mothers were the main
153 informants. It should be noted researchers mostly used the term 'parent(s)' to present their
154 findings therefore, the research reviewed did not consistently identify impacts specific to
155 either mother or father.

156
157
158 **TITLE What is the psycho-social impact on parent caregivers of young people living**
159 **with muscular dystrophy?**

160
161 **The impact on parents of caring for children with DMD, changed over time, as the**
162 **child's dependency increased as they moved from childhood to adulthood. The impacts**
163 **encompassed not only psycho-social aspects of their lives but also physical and**
164 **financial (*Living with DMD*). In addition, communicating information about DMD**
165 **emerged as a significant issue (*Knowing and telling*) along with managing transitional**
166 **care (*Transitioning*).**

167 168 **4.1. *Living with DMD***

169 **4.1.1. Psycho-social impacts**

170 Parents reported having feelings of loss, sadness and depression [6, 11] as they lived
171 with their child's condition but identified their main psychological issue as their distress
172 and worry for the future [14,15]. These parents experienced significantly greater stress
173 than parents of healthy children and where their child also had difficulties in social
174 interactions, this further increased stress levels [16]. Generally, parents felt they had
175 coping mechanisms for day-to-day living but reported struggling, at times, with both
176 interventions and behavioural changes [14]. In addition, parents not only worried for
177 their affected child but also for the negative influence DMD had on the psychological well-
178 being and social life of the siblings [11,15].

179 Although continually living with stress and worries for their child and family, peaks
180 in parental stress levels were experienced at life-changing moments, notably the time of
181 diagnosis, the point where disease progression rendered their sons immobile, when a
182 powered wheelchair or non-invasive ventilation (NIV) became necessary and at the death
183 of peers [12,16,17]. These major events marked out disease progression with parents
184 identifying their child's loss of ambulation as one of the most difficult challenges to cope
185 with emotionally [16]. As parents struggled with the psychological, physical and, for

186 some, financial impacts of their child's disease, they often disengaged from their own
187 hobbies and social activities [15].

188 Despite the many negative psychological impacts of being a parent of a child with
189 DMD, parents repeatedly identified their experiences as positive [11,15,18,19]. Those
190 most positive about their situation, were long term caregivers and those who viewed their
191 child as being both sensitive and talented: very few mothers reported any negative
192 feelings about assisting their child [18]. The parents' experiences often changed their
193 personal life values and increased their strength and courage in facing adversity [11].
194 Parents were themselves aging as their child's dependence was increasing but they felt
195 increased confidence from having raised their children and satisfaction with their work
196 as caregivers [19]. In contrast, some parents who had developed health problems
197 themselves (sight issues, back problems and hypertension) had concerns about not being
198 as able to care for their child as previously [19].

199 4.1.2. Physical and financial impacts

200 Home management of their child's care was a demanding role for parents and this
201 physical care burden was perceived as greatest where the child had suffered with the
202 disease longer, had lower functional ability and was more dependent on caregivers
203 [11,15]. The demands on parents were extensive with many reporting night-time
204 waking to give care and respond to equipment alarms, especially in the later disease
205 stages due to the child's immobility and need for non-invasive ventilation (NIV).
206 Malfunction or dislodgement of NIV can be fatal for the child should the care giver not
207 intervene and these nighttime care demands, negatively impacted the quantity and
208 quality of sleep for parental caregivers, with those less experienced being most adversely
209 affected [17]. Perception of the extent of the care burden was linked with parental access
210 to social contact and support from friends, family and professionals especially in
211 emergency situations [11, 15].

212
213 The nature of the disease meant there was a growing physical dependency on parents
214 as the child matured and an increasing financial burden for some families [5,17]. As their
215 child grew into a young man, parents anxiously anticipated their own ageing, retirement
216 and the changes in family relationships and structures as their other children grew up and
217 had families [19]. For parents these events coincided with increasing care needs and
218 financial burden for them alongside the loss of their own primary caregiver role [19].

219
220 Parents admitted harbouring some regrets for the life constraints that DMD had
221 imposed on them, some of which were financial [19]. Some studies have demonstrated
222 that families with a child with DMD have a lower than national median income with many
223 costs associated with care provision resulting in substantial economic burden for families
224 [5,20] and in low socio-economic countries the impact on families of the disease was even
225 greater [12]. Economic worries were real, and families found it difficult to escape poverty
226 or even think about how to increase their income [19].

227 228 4.2. *Knowing and telling*

229 This theme recognises the significance of the moment when parents were first told of
230 their child's diagnosis. It was at this stage that parents were told of their child's prognosis
231 with this genetic, degenerative disease and mothers first found out of their own potential
232 carrier risk and that of their daughters [21].

233 Mothers felt responsible for telling their daughters about their carrier risk and
234 imparted critical pieces of knowledge to them at key developmental stages, for example,
235 starting high school, turning sixteen or being in a serious relationship [21]. This lengthy,
236 complex process allowed daughters to assimilate the knowledge as they matured. Six
237 levels of disclosure of information were identified: condition; genetics; carrier risk; carrier
238 test requested; reproductive options and carrier testing; life expectancy. Mothers who

239 told their daughters the most information, often did this through unplanned
240 conversations as they responded to questioning. Lack of knowledge about advanced
241 genetic reproduction options and beliefs about timing of discussions acted as key
242 communication barriers for mothers. Similarly, reduced life expectancy was considered
243 too difficult a topic to talk about so was generally avoided [21].
244

245 It was proposed that following diagnosis parents would benefit from having written
246 information about DMD which should include key facts, a summary of reproductive
247 options and advice about how to tell daughters this critical information. In addition,
248 genetic counselling and psychological support for both daughters and mothers may help
249 to mitigate the long-lasting guilt and blame often felt by carriers [12,21].

250 Discussion of potential carrier status with daughters posed specific challenges but
251 equally parents struggled about talking to their sons about their disease and its
252 progression [12]. Parents reported that their own lack of knowledge and understanding
253 about DMD, its progression, advances in treatment and access to supportive services often
254 prevented them effectively communicating with their child [12]. One parent had told his
255 son he had 'weak muscles' when his child was upset, he could no longer run or play, other
256 parents avoided conversations about disease progression altogether [12]. Parents wanted
257 more knowledge, so they were better equipped for these conversations [12] and even at
258 transition to adult services parents still highlighted their need to understand DMD and be
259 up to date, in readiness to be a 'back-up' carer [22]. These findings demonstrate that
260 throughout their journey, parents have information and support needs that are unmet,
261 leaving them struggling to communicate and help their children who are affected by
262 DMD.
263

264 4.3. Transitioning

265 Increased life expectancy in young people with DMD, due to interventions
266 such as ventilation, is allowing caregivers to be aspirational about their child's
267 future. Transition has been described as not only moving from paediatric to adult
268 care, from childhood to adulthood but also in terms of disease progression.
269 During teenage years young people begin to plan their futures including
270 continuing education, entering the work of work, and expanding their social
271 relationships. For boys with muscular dystrophy trying to emerge into adulthood
272 occurs at a time when their physical dependence on others is increasing.

273 The age of transition occurs much later than the normal population [22]. The
274 experience of young people with disabilities is one where they are falling behind
275 their peers in fulfilling adult social roles due to reduced opportunities, lack of
276 expectation and overprotectiveness from those involved in their care. **For**
277 **successful family functioning, during transition, balance needs to be achieved**
278 **between the needs of the young person and those of the rest of the family [23].**

279 **Parents themselves need to change their behaviours during transition,**
280 **moving from a 'manager' role to one of 'consultant' as their son matured [24,25]**
281 **but remaining a strong influence [22,26].** Having been a primary caregiver for a
282 prolonged period brings with its additional challenges [19]. Entrusting their son's
283 care to aides was difficult as parents fundamentally believed that no-one except
284 them could always put him first. Parents became deeply concerned about their
285 son's future as they became older themselves [22]. At this time of transition, the
286 majority were dependent on mechanical ventilation and parents lived with a
287 sense of impending crisis knowing their son's life was reliant on the mechanical
288 ventilator and care. Parents acknowledged that whilst trying not to meddle they
289 knew they swayed their son's decisions because of their own anxieties.

290 It is possible that, as DMD progresses, parents may feel overwhelmed by their
291 caring role, and too exhausted to be involved in social activities. This situation
292 may lead to a vicious cycle of events where parents gradually reduce their social
293 engagement to cope but in fact this may expose them to greater burden as time
294 progresses, with further social withdrawal impacting on transition [15,21].

295 The role of parents in transition, is multifaceted. Whilst maintaining their role
296 as a “lifeline” they need to adapt by changing parenting and caring behaviours
297 [22]. There is a requirement to increase the flexibility of family boundaries to
298 permit development of children’s independence. Parents need to develop new
299 adult relationships with their children [23].

300 **What therapeutic interventions are employed to meet the psycho-social needs of parent** 301 **caregivers of young people living with muscular dystrophy?** 302

303 A limited number of the reviewed papers outlined potential therapeutic interventions
304 aimed at addressing psycho-social needs of parent care-givers. The key theme, *Building*
305 *resilience*, underpins interventions identified in the literature which focussed on
306 *adapting, well-being, socialising and escaping.*

307 *4.4. Building resilience*

308 *4.4.1 Adapting*

309 Resilience is not just the ability to bounce back from adversity but the process
310 of adapting [27]. There is a need to foster the mother’s resilience using psycho-
311 social interventions aimed at improving acceptance by identifying the positive
312 aspects of living with DMD rather than just the burden and deficit [11,28,29].

313 Psycho-social support should start when the child is young and those
314 involved in caring for families should assess unmet needs. Being proactive in
315 identifying the need for help and understanding the families fears and
316 uncertainty also helps in the identification of resources needed to prioritise and
317 customise interventions building on family strengths [30].
318

319 *4.4.2 Wellbeing*

320 Parental health is seen as a necessity for family adaptation [11,29,31], however,
321 the requirements for good parental health are varied. The family environment
322 can contribute to or mitigate burden [19,30]. Family and partner support are
323 seen as important [30]. It has been found that an intact family structure may
324 influence family hardiness and the provision of emotional support [30,31,32]. If
325 the parent is single, or the child not of school age there is **greater need for social**
326 **and professional support to manage care and minimise burden [11].**

327 **Parents of children with DMD experience higher levels of stress than other**
328 **parents [13,29] and it is reported that distressed or depressed parents may**
329 **become frustrated and see their children as more of a burden [29]. Where stress**
330 **is related to their child, or problem behaviours such as clinginess and poor**
331 **socialisation skills, interactions between mother and child also become more**
332 **stressful [13]. This correlation between parental stress and psycho-social**

333 adjustment can lead to a decline in good parenting skills and poorer coping
334 mechanisms [29].

335 Good parental health and management of stress is reliant on responsive
336 community services [6,11,31] and supportive health professionals [30]. Attendance
337 at support groups, where parents can discuss their fears and anxieties, gain advice
338 on resources and expand their knowledge, is seen to reduce levels of stress and
339 aid good psycho-social adjustment [29].
340

341 4.4.3 Socialising

342 Creating opportunities for socialization is an important intervention, not just for the
343 young person but the parent [6,24,25]. Social support is frequently used by caregivers as
344 a coping strategy [25] and whilst it can be challenging the benefits are clear [24]. Parents
345 found that engaging in support groups gave them access to practical advice, emotional
346 support and helped them to understand their child's condition better [25]. Sharing and
347 improving knowledge is linked with active coping [8] giving parents power and the voice
348 to advocate for their child [6].

349 Access to social support and the opportunity to socialise outside of the home can allow
350 the caregiver to escape (24) or get away (8) from caregiver responsibilities. This becomes
351 more important as DMD progresses as there is a potential for caregivers to reduce social
352 engagement to cope with caring responsibilities and thus further increase burden. To
353 avoid this vicious cycle of events there is a need to build upon existing social supports to
354 improve the psychosocial outcomes for families coping with the effects of DMD.

355 4.4.4 Escaping

356 There was acknowledgement that parents could benefit from having some time-out
357 from their role and responsibilities. Escaping the ongoing daily pressures could provide
358 some well needed relief for parents. So, pain employment may not only have financial
359 benefits but also could be a means of escaping care burden even if only for short periods
360 of time [11, 31]. This time out is important.

361 Parents also identify exercise and self-care activities as being necessary for their overall
362 health [30] however many neglect hobbies [11] and spend little time on social activities
363 and rest [32]. Over time, as their child matured and became more dependent, the
364 parents' neglect of their own well-being often increased [15] and their quality of life
365 deteriorated [33]. Respite care can improve caregiver burden [31] but uptake is low [29].
366

367 **What factors influence accessibility of psycho-social interventions for these caregivers?**

368

369 Accessibility of psycho-social interventions for caregivers was not a central focus of the
370 studies, therefore, this research question was not fully addressed, indicating a need for
371 further research in this area. Researchers noted variation in the level of professional and
372 social support for parents within their studies but factors influencing this situation were
373 not explored. For example, it was noted that uptake of respite care is low [29] but the
374 reasons for this are not clearly established. It is however suggested that it could be 'too
375 time consuming to organise' or due to maternal anxiety about relinquishing care [30]. It
376 is possible there are many issues underlying the decision making in accessing respite
377 services and therefore to fully understand this, future research needs to concentrate on
378 uncovering the potentially complex influencing factors.

379
380 Similarly, there is recognition that good parental health and management of stress is
381 reliant on responsive community services [6,11,31] and supportive health professionals
382 [30]. However, some families struggled to access resources and had to fight for services
383 identifying a lack of joined up thinking as a barrier to consistent care [6]. Without a fuller
384 understanding of these critical access issues it is difficult to draw any conclusions
385 regarding factors influencing this situation.
386
387

388 5. Discussion

389
390 The review findings demonstrated the impact on parents caring for children
391 with DMD changed over time and encompassed psycho-social, physical and
392 financial aspects of their lives. Parents lived with continuing levels of stress and
393 worry which peaked at diagnosis and with each milestone in their child's disease
394 progression. These key markers of life changing moments can be predicted and
395 include their child's loss of mobility, need for enteral feeding, need for NIV, transition
396 to adulthood and when their friends die. This heavy care burden for parents and impact
397 on parents' lives has previously been recognised in the literature [23]. However, this
398 review extends our understanding of the pressures on parents particularly at identifiable,
399 predictable life-changing moments and this knowledge is critical to development of
400 improved parental support and information provision.
401

402 What is evidenced within the review is that some families have more help and
403 support around them. Social circumstances, availability of professional and family
404 support and information about DMD influenced family experiences of living with the
405 condition. Several studies reported unmet parental needs changing throughout the child's
406 disease trajectory, impacting upon communication, education and coping. The parental
407 dilemmas in communicating with their child about disease progression and prognosis,
408 uncovered in the review, echoes previous findings about disclosures to children with a
409 cancer diagnosis [34,35,36,37]. Adoption of suggested communication improvements
410 from cancer care research could be equally beneficial for parents of children with DMD,
411 as the role of information gatekeeper seems common to both parent groups.
412

413 Gibson et al [37] found that there was often a mismatch between professionals'
414 communication and parental information needs within individual changing
415 circumstances. So, it may be useful within the context of DMD for professionals to aim to
416 better match their communications with parental needs, as they too experience altering
417 circumstances as their child's disease progresses.

418 Despite the many demands on parents some were still able to find positives in their
419 experiences. They articulated satisfaction in their caregiving having raised their child and
420 development of confidence, personal strength and courage in adversity were reported.
421 No mothers identified negative feelings in caring for their child. These positive
422 evaluations of parenthood whilst they managed their heavy care burden mirrors existing
423 research findings from parents of adult sons with DMD [38]. Lazarus and Folkman's
424 Transactional Model of Stress [39] may offer explanation for these findings, in that parents
425 with adequate resources to meet the demands placed on them may then see positives in
426 their experiences.

427 Findings from the review suggested that transition into adulthood brings additional
428 challenges for the young person and their parents. The timing of transition occurs later
429 for the young person with DMD and requires both parties to adopt different behaviours
430 to facilitate role change. The parents need to 'let go' and by doing so the young person can
431 learn to direct their own health care. Transition into adulthood brings additional
432 challenges for the young person and their parents. One important aspect of this, suggested
433 by Sonneveld [40] is recognising that the changes in responsibility are managed gradually.

434 As parents begin the process of ‘letting go’ they will feel a loss of control [27,41], however
435 they can be reassured that to manage their transition successfully the young person will
436 need to use their parent as a ‘safety net’. They are not abandoning their child but allowing
437 them to grow using their new role as facilitator.

438
439 It is interesting to note that analysis of the reviewed papers demonstrated a lack of
440 reference to palliative care despite DMD being a progressive disorder leading to reduced
441 life expectancy. This could be because they view a changing approach to care as a natural
442 trajectory and may not necessarily understand terminology. Terminal care, palliative care,
443 supportive care, hospice care and end of life care are all terms that may become confused,
444 feared and misunderstood [42] or avoided. The lack of consensus and understanding of
445 the constantly changing nomenclature can undermine care provision and confuse the way
446 care is delivered to people who are most vulnerable at the end of their lives [43,44]. This
447 was not a focus of the review but is worthy of further exploration as to how the subject is
448 approached with parents of children with DMD.
449

450 6. Conclusion

451 This integrative review highlights areas for improvement in meeting the parent’s
452 psycho-social needs, critical to improving parents’ experiences is anticipating their needs
453 in advance. The review has identified major stress points where parents need additional
454 support and information to help them to adapt during challenging periods. Armed with
455 this knowledge health care professionals, utilising a needs assessment approach, can
456 target interventions in a timely manner to promote adaptation and build resilience. It is
457 proposed that by more effectively supporting the parent(s) this will potentially have
458 positive effects on family functioning.

459 Medical advances have changed the landscape of care and have allowed parents to
460 become more aspirational in planning for the future. **The advent of revolutionary new
461 drug therapies should bring dramatic impacts for families. This was not an aspect covered
462 in this review as the children had not yet benefitted from these new therapies.**

463 What is needed is more focus on helping parents adapt to their growing child and
464 change their role back from caregiver to parent, allowing the young people to also be
465 aspirational about their future.

466 Evidence suggests that there is benefit in challenging perspectives so that the focus
467 is not always on deficit and burden but on positivity and opportunities. The nature of the
468 disease has the potential to isolate and reduce social skill development essential for
469 transition to adulthood. As interactive situations with peers and adult role models with
470 DMD is seen to have positive impacts on both young people and parents.

471 It is proposed that future investment should focus on anticipating family need,
472 targeting intervention cognisant of predictable stress points and building resilience
473 through social community. Parents may then be better positioned to support their child
474 in looking forward.
475

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483 References

1. Whitemore, R; Knafl, K. (2005) The integrative review: updated methodology. *Journal of Advanced Nursing* Vol. 52 (5), pp. 546-53. <https://10.1111/j.1365-2648.2005.03621.x>
2. Waldrop A.N.J. (2020) Overview of gene therapy in spinal muscular atrophy and Duchenne Muscular Dystrophy. *Pediatric Pulmonology*. Published online Sept, 4 2020. Doi: 10.1002/ppul.25055
3. Verma A. (2018) Recent advances in Antisense Oligonucleotide Therapy in Genetic Neuromuscular Diseases. *Annals of Indian Academy of Neurology* 21(1), pp3-8 doi: 10.4103/aian.AIAN_298_17
4. Greener M. (2020) Making sense of antisense oligonucleotide therapy. *Prescriber*, vol.31 (6), pp15-18.
5. Landfeldt, E; Lindgren, P; Bell, C; F; Guglieri, M; Straub, V. et al. (2016) Health-related quality of life in patients with Duchenne muscular dystrophy: a multinational, cross-sectional study. *Developmental Medicine and Child Neurology* Vol. 58 (5), pp. 508-515. <https://doi.org/10.1111/dmcn.12938>
6. Hoskin, J. (2017) Taking charge and letting go: exploring ways a transition to adulthood project for teenagers with Duchenne Muscular dystrophy has supported parents to prepare for the future. *British Journal of Special Education*, Vol. 44 (2), pp. 165-185 <https://doi.org/10.1111/1467-8578.12173>
7. Hopia, H; Latvala, E; Liimatainen L. (2016) Reviewing the methodology of an integrative review *Scandinavian Journal of Caring Sciences* Vol.3 (4) pp. 57-70 <https://doi.org/10.1111/scs.12327>
8. Dwyer, P.A. (2020) Analysis and Synthesis, A Step-by-Step Guide to Conducting an Integrative Review, <https://doi.org/10.1007/978-3-030-37504-1>
9. Rothstein, H; Hopewell, S. (2009). *Grey literature*. In H. Cooper, L. V. Hedges, and J. C. Valentine (Eds.), *The Handbook of Research Synthesis and Meta-analysis*. Russell Sage Foundation.
10. Lson, CA. (2013) Using the grey literature to enhance research and practice in continuing education for health professionals. *Journal Continuing Education Health Professional* Vol 33 (1) pp. 1– 3. <https://doi.org/10.1002/chp.21159>
11. Magliano, L; Politano, L. (2016) Family context in muscular dystrophies: psychosocial aspects and social integration. *Acta Myologica: Myopathies and Cardiomyopathies* Vol. 35 (2), pp. 96-99.
12. Thomas, PT; Rajaram, P; Nalini, A. (2014) Psychosocial challenges in family caregiving with children suffering from Duchenne muscular dystrophy. *Health and Social Work* Vol. 39 (3), pp. 144-52. <https://doi.org/10.1093/hsw/hlu027>
13. Nereo, N; Fee, R; Hinton, V. (2003) Parental Stress in Mothers of Boys with Duchenne Muscular Dystrophy. *Journal of Paediatric Psychology* Vol 28 (7) pp. 473-484. <https://doi.org/10.1093/pepsy/jsg038>
14. Ravens- Sieberer, U; Erhart, M; Wille, N; Wetzel R; Nickel J; et al. (2006) Generic Health Related Quality of Life Assessment in Children and Adolescents *Pharmacoeconomics* Vol 24 (12) pp. 1199-1220. <https://doi.org/10.1177/0269472706024121199>
15. Magliano, L; D'Angelo, MG; Vita, G; Pane, M; D'Amico, A; Balottin, U. et al. (2014) Psychological and practical difficulties among parents and healthy siblings of children with Duchenne vs. Becker muscular dystrophy: an Italian comparative study. *Acta Myologica: Myopathies and Cardiomyopathies* Vol. 33 (3), pp. 136-43.
16. Bray, P; Bundy, AC; Ryan, MM; North, KN; Burns, J. (2011) Health status of boys with Duchenne muscular dystrophy: A parent's perspective. *Journal of Paediatrics and Child Health* Vol 47 (8) pp. 557-562. <https://doi.org/10.1111/j.1440.1754.2011.02022.x>
17. Nozoe, KT; Polesel, DN; Moreira, GA; Pires, GN; Akamine, RT; et al. (2016) Sleep quality of mother-caregivers of Duchenne muscular dystrophy patients. *Sleep and Breathing* Vol. 20 (1), pp. 129-34. <https://doi.org/10.1007/s11325-015-1196-9>
18. Magliano, L; Patalano, M; Sagliocchi, A; Scutifero, M; Zaccaro, A. (2014) 'I have got something positive out of this situation': psychological benefits of caregiving in relatives of young people with muscular dystrophy, *Journal of Neurology* Vol. 261 (1), pp. 188-95. <https://doi.org/10.1007/500415-013-7176-8>
19. Yamaguchi, M; Sonoda, E; Suzuki, M. (2019) The experience of parents of adult sons with Duchenne muscular dystrophy regarding their prolonged roles as primary caregivers: a serial qualitative study. *Disability and Rehabilitation* Vol. 41 (7), pp 746-752. <https://doi.org/10.1080/09638288.2017.1408148>

- 531 20. Magliano, L; Scutifero, M; Patalano, M; Sagliocchi, A; Zaccaro, A; et al. (2017) Integrated care of muscular
532 dystrophies in Italy. Part 2. Psychological treatments, social and welfare support, and financial costs. *Acta*
533 *Myologica: Myopathies and Cardiomyopathies* Vol. 36 (2), pp. 41-45.
- 534 21. Goldman, A; Metcalfe, A; MacLeod, R. (2018) The Process of Disclosure: Mothers' Experiences of
535 Communicating X-Linked Carrier Risk Information to At-Risk Daughters. *Journal of Genetic Counselling*, Vol.
536 27 (5), pp. 1265-1274. <https://doi.org/10.1007/s10897-018-0251-7>
- 537 22. Yamaguchi, M; Suzuki, M (2015) Becoming a back-up carer: parenting sons with Duchenne muscular
538 dystrophy transitioning into adulthood. *Neuromuscular Disorders* Vol. 25 (1), pp. 85-93.
539 <https://doi.org/10.1016/j.nmd.2014.09.001>
- 540 23. Waldboth V; Patch C; Mahrer-Imhof R; Metcalfe A. (2016) Living a normal life in an extraordinary way: A
541 systematic review investigating experiences of families of young people's transition into adulthood when
542 affected by a genetic and chronic childhood condition. *International Journal of Nursing Studies* Vol. 62, pp. 44-
543 59. <https://doi.org/10.1016/j.ijnurstu.2016.07.007>
- 544 24. Parkyn, H, Coveney, J. (2013) An exploration of the value of social interaction in a boys' group for adolescents
545 with muscular dystrophy. *Child: Care, Health and Development* Vol 39 (1) pp. 81-89.
546 <https://doi.org/10.1111/j.1365-2214.2011.01353.x>
- 547 25. Kingsnorth, S; Rudzik, AEF; King, G; McPherson, AC. (2019) Residential immersive life skills programs for
548 youth with disabilities: a case study of youth developmental trajectories of personal growth and caregiver
549 perspectives. *BMC Pediatrics*, Vol. 19 (1), pp. 413. <https://doi.org/10.1186/s12887-019-1793-z>
- 550 26. Shirazipour, CH; Latimer-Cheung, AE; Arbour-Nicitopoulos, KP. (2015) An exploratory qualitative
551 investigation of psychosocial determinants of parental decisions to support sport participation for youth with
552 a mobility impairment. *Research in Developmental Disabilities* Vol. 45-46, pp. 400-10.
553 <https://doi.org/10.1016/j.ridd.2015.08.001>
- 554 27. Westwood, A; Langerak, N; and Fiegggen, G. (2014) 'Transition from child- to adult orientated care for children
555 with long-term health conditions: A process, not an event.' *Continuing Medical Education*, Vol. 104 (4), pp.310-
556 313. <https://doi:10.7196/SAMJ.8201>
- 557 28. Peay, HL; Meiser, B; Kinnett, K; Furlong, P; Porter, K. et al. (2016) Mothers' psychological adaptation to
558 Duchenne/Becker muscular dystrophy. *European Journal of Human Genetics*: Vol. 24 (5), pp. 633-7
559 <https://doi:10.1038/ejhg.2015.189>
- 560 29. Gocheva, V; Schmidt, S; Orsini, AL; Hafner, P; Schaedelin, S; et al. (2019) Psychosocial adjustment and
561 parental stress in Duchenne Muscular Dystrophy. *European Journal of Paediatric Neurology* Vol. 23 (6), pp. 832-
562 841. <https://doi.org/10.1016/j.ejpn.2019.09.008>
- 563 30. Peay, H.L; Meiser, B; Kinnett, K; Tibben, A. (2018) Psychosocial needs and facilitators of mothers caring for
564 children with Duchenne/Becker muscular dystrophy. *Journal of Genetic Counselling*, Vol. 27 (1), pp. 197-203.
565 <https://doi:10.1007/s10897-01700141-4>
- 566 31. Chen, JY; Clark, MJ. (2010) Family resources and parental health in families of children with Duchenne
567 Muscular dystrophy. *The Journal of Nursing Research*: Vol. 18 (4), pp. 239-48.
568 <https://doi.org/10.1097/JNR.0b013e3181f3e37b>
- 569 32. Suk, Kyung S; Baek, JH; Park, JO; Kim, HS; Lee, HM; et al. (2015) Postoperative quality of life in patients with
570 progressive neuromuscular scoliosis and their parents. *The Spine Journal* Vol. 15 (3), pp. 446-453.
571 <https://doi.org/10.1016/j.spinee.2014.09.030>
- 572 33. Jones, KM; O'Grady, G; Rodrigues, MJ; Ranta, A; Roxburgh RH; et al. (2018) MD-PREV study group, Impacts
573 of Children Living with Genetic Muscle Disorders and their Parents – Findings form a Population-Based
574 Study. *Journal of Neuromuscular Diseases* Vol. 5 (3), pp. 341-352. <https://doi.org/10.3233/JND-170287>
- 575 34. Mack, J; Wolfe, J; Grier, HE; Cleary, PD; Weeks, JC. (2006) Communication about prognosis between parents
576 and physicians of children with cancer: Parent's preferences and the impact of prognostic information.
577 *Journal of Clinical Oncology*, Vol 24 (33), pp. 5265-5270. <https://doi.org/10.1200/JCO.2006.06.5326>

- 578 35. Mack, JW; Wolfe, J; Cook, EF; Grier, HE; Cleary P. et. al (2011) Parents' roles in decision making for children
579 with cancer in the first year of cancer treatment. *Journal of Clinical Oncology*, Vol 29 (15), pp. 2085-2090.
580 <https://doi.org/10.1200/JCO.2010.32.0507>
- 581 36. Sisk, BA; Bluebond-Langner, M; Wiener, L; Mack, J., and Wolfe, J. (2016) Prognostic disclosures to children: a
582 historical perspective. *Pediatrics*, Vol. 138 (3). <https://dx.doi.org/10.1542%2Fpeds.2016-1278>
- 583 37. Gibson, F; Kumpunen, S; Bryan, G; and Forbat, L. (2018) Insights from parents of a child with leukaemia and
584 healthcare professionals about sharing illness and treatment information: A qualitative research study.
585 *International Journal of Nursing Studies*, Vol. 83, 91-102.DOI: [10.1016/j.ijnurstu.2018.04.008](https://doi.org/10.1016/j.ijnurstu.2018.04.008)
- 586 38. Pangalila, RF; van den Bos, EA; Geertrudis, AM; Stam, HJ; van Excel NJ. et al. (2012) Subjective caregiver
587 burden of parents of adults with Duchenne muscular dystrophy. *Disability and Rehabilitation* Vol.34 (12), pp.
588 988-996. <https://doi.org/10.3109/09638288.2011.628738>
- 589 39. Lazarus, R; Folkman, S. (1984) *Stress, Appraisal and Coping*. New York: Springer; 1984
- 590 40. Sonneveld, H; Strating, M; Staa, ALV; and Nieboer, AP. (2011) 'Gaps in transitional care: what are the
591 perceptions of adolescents, parents and providers?' *Child: Care, Health and Development*. Vol. 39 (1), pp.69-80.
592 <http://doi.org/10.1111/j.1365-2214.2011.01354.x>
- 593 41. Schlucter, J. (2014) 'Patient and Family-Centred Transitions from Pediatric to Adult Care' *Paediatric Nursing*
594 Vol 40 (6), pp.307-310.
- 595 42. Gott M. Frey R. Robinson J. et al. (2013) The Nature of and Reasons for "inappropriate" Hospitalisation
596 Among Patients with Palliative Care Needs: A Qualitative Exploration Of the Views of Generalist Palliative
597 Care Providers. *Palliative Medicine* 0 (0) p1-10.
- 598 43. Mannix K. (2017) *With the End In Mind Dying, Death and Wisdom in an Age of Denial*. London. William
599 Collins.
- 600 44. Pastrana T. Junger S. Ostgathe C. et al. (2008) A Matter of Definition-Key Elements Identified in a Discourse
601 Analysis of Definitions of Palliative Care. *Palliative Medicine* 22 (3) p222-232.
602
603