Decision-making and future planning for children with life-limiting conditions: A qualitative systematic review and thematic synthesis

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Abstract

Background: In the last decade the number of children with life-limiting and life-threatening conditions in England has almost doubled and it is estimated that worldwide there are 1.2 million children with palliative care needs. Families and professionals caring for children with life-limiting conditions are likely to face a number of difficult treatment decisions and develop plans for future care over the course of the child's life, but little is known about the process by which these decisions and plans are made.

Methods: The purpose of this review is to synthesise findings from qualitative research that has investigated decision making and future planning for children with life-limiting conditions. A systematic search of six online databases was conducted and identified 887 papers for review; 5 papers were selected for inclusion, using pre-defined criteria. Reference list searching and contacting authors identified a further 4 papers for inclusion.

Results: Results sections of the papers were coded and synthesised into themes. Nineteen descriptive themes were identified and these were further synthesised into four analytical themes. Analytical themes were 'decision factors', 'family factors', 'relational factors' and 'system factors'.

Conclusions: Review findings indicate that decision making and future planning is difficult and needs to be individualised for each family. However, deficits in understanding the dynamic, relational and contextual aspects of decision making remain and require further research.

Background

The prevalence of children with life-limiting and life-threatening conditions in England is approximately 40,000 and this number is increasing (Fraser et al., 2012). Worldwide it is estimated that there are 1.2 million children with life-limiting and life-threatening conditions (Worldwide Palliative Care Alliance, 2014). "Life-limiting conditions are those

for which there is no reasonable hope of cure and from which, ultimately, the child will die. Life-threatening conditions are those for which curative treatment may be feasible but can fail" (Together for Short Lives, 2014, p.1). There are four categories of life-limiting and life-threatening conditions (ACT and RCPCH, 1997, see Table 1). Within this paper, the term life-threatening condition will be used to refer to category 1 and the term life-limiting condition will be used to refer to categories 2-4. Life-threatening and life-limiting conditions pose different challenges for families and professionals in relation to treatment and decision making. The literature suggests that a number of significant decisions may need to be considered for children with life-limiting conditions over the course of their life, including commencing artificial feeding, assisted ventilation and surgical intervention (Carroll et al., 2012, Guerriere et al., 2003). Such decisions are complex and influenced by more than simple exchange of information (Carroll et al., 2012). Some literature suggests that families of children with disabilities experience emotional and physical exhaustion (Heiman, 2002, Green, 2007, Kratz et al., 2009), which may compound the difficulties of decision making.

Category	Descriptor
1	Life-threatening conditions for which curative treatment may be feasible but can fail.
2	Conditions where premature death is inevitable. There may be long periods of intensive treatment aimed at prolonging life and allowing participation in normal activities.
3	Progressive conditions without curative treatment options. Treatment is exclusively palliative and may commonly extend over many years.
4	Irreversible but non-progressive conditions causing severe disability, leading to susceptibility to health complications and likelihood of premature death.

Table 1: Categories of life-limiting and life-threatening conditions (ACT and RCPCH, 1997)

Few studies have investigated decision making or future planning specifically for children with life-limiting conditions. Within this paper decision making will refer to choices made regarding current treatment and care options, or future care plans and will focus not on the decision made, but on the process of making it. Much of the literature regarding decision making within paediatrics has been conducted in the field of paediatric oncology (Price et al., 2012). Although findings from this body of literature may help in developing an understanding of the factors influencing decision making, the results may not be directly applicable to children with non-curable life-limiting conditions. Given the dearth of

research investigating decision making in this context, this review focussed on qualitative research, as it enables a deeper understanding of the context (Miles and Huberman, 1994) within which a phenomena occurs and an enhanced understanding of the phenomena itself (Sofaer, 1999). Reviewing the qualitative research therefore, will help to highlight the processual and contextual factors involved in decision making. Therefore, this paper will provide a review and synthesis of the qualitative research that specifically focusses on decision making and future planning, solely for children with life-limiting conditions within categories 2-4 (Table 1).

Methodology

The thematic synthesis of qualitative research aims to provide a re-interpretation of previous findings, such that the conclusions of the review go beyond the conclusions developed within the individual research studies (Thomas and Harden, 2008, Sandelowski and Barroso, 2006). Thematic Synthesis offers an approach for researchers to make practical recommendations to practitioners and policy makers (Barnett-Page and Thomas, 2009). The data driven process involves extracting and coding the text, developing descriptive themes and sub-themes, and generating analytical themes (Thomas and Harden, 2008). Inferences are made regarding the relationships between the descriptive themes and research questions, resulting in abstract analytical themes.

Eligibility Criteria

The eligibility criteria for inclusion in the review are outlined in **Table 2**.

Inclusion Criteria
• The study must focus on children (>28 days old) and young people with a life-limiting illness within categories 2-4 (see Table 1).
• The study must involve health care professionals, family members, 'significant others' or the child who make decisions about the child's care.
• The study must be qualitative in design or have a qualitative component. It will consider studies that focus on qualitative data including, but not limited to, designs such as phenomenology, grounded theory, ethnography and action research. Descriptive qualitative studies that describe the experience or describe the effects of the experience will also be considered.
• Studies recruiting professionals from ICU's if the focus of investigation is children with pre-existing life-limiting illness.

Exclusion criteria
Any studies: • including children or young people with life-threatening illness.   • where families or professionals are recruited from ICU's, unless the focus is specifically on decision making for children with life-limiting conditions.   • recruiting families or professionals from neonatal ICU as many of the participants will be discussing children under the age of 28 days and therefore outside the criteria of the study   • using purely quantitative methods.

Table 2: Thematic synthesis eligibility criteria

To maximise the opportunity for inclusion, studies were included if they investigated decision making for people with life-limiting conditions, as long as the study involved some life-limited children (28 days-18 years old) in the sample. The decision to include studies which included adults in their sample has implications for the results of the synthesis. However, these were weighed up against the lack of research in the area and the need to synthesise the existing knowledge in the field and highlight gaps for further investigation.

Searching for literature

Within this synthesis a systematic approach to searching was taken to maximise and balance specificity and sensitivity. In order to identify the relevant literature, various bibliographic databases, covering the fields of health care and psychology, were searched; these were CINAHL, Medline, Embase and PSYCHInfo. Indexed terms and keywords were used to find papers relevant to the review question. In order to find any relevant literature outside of the health field, a free text search was conducted in ProQuest at the advice of a librarian. Additionally, EThOS (E-Theses Online Service) was searched using the words "life limiting" AND "child", in order to identify unpublished doctoral theses. The initial electronic search was conducted in December 2014 and has been updated periodically until April 2016. The search strategy used in one of the databases (Medline) is provided below (Figure 1). Authors of relevant conference abstracts were also contacted to identify if they had articles presenting the findings from their research or for unpublished work.

1. emergency care planning.mp.
2. emergency healthcare plan.mp.
3. exp Patient Care Planning/
4. patient care conference.mp.
5. exp Advance Care Planning/ or exp Advance Directives/
6. exp Advance Directives/
7. advance medical plan.mp.
8. advance healthcare plan.mp.
9. advance health care plan.mp.
10. exp Euthanasia, Passive/
11. withhold* treatment.mp. [mp=title, abstract, original title, name of substance word, subject heading word, keyword heading word, protocol supplementary concept word, rare disease supplementary concept word, unique identifier]
12. withdraw* treatment.mp. [mp=title, abstract, original title, name of substance word, subject heading word, keyword heading word, protocol supplementary concept word, rare disease supplementary concept word, unique identifier]
13. exp Withholding Treatment/
14. exp Resuscitation Orders/
15. medical order.mp.
16. exp Patient Care Planning/
17. goals of care.mp.
18. care goals.mp.
19. values history.mp.
20. values clarification.mp.
21. (goals and objectives).mp. [mp=title, abstract, original title, name of substance word, subject heading word, keyword heading word, protocol supplementary concept word, rare disease supplementary concept word, unique identifier]
22. exp Palliative Care/
23. exp Terminal Care/
24. 1 or 2 or 3 or 4 or 5 or 6 or 7 or 8 or 9 or 10 or 11 or 12 or 13 or 14 or 15 or 16 or 17 or 18 or 19 or 20 or 21 or 22 or 23
25. exp Decision Making/
26. exp Decision Support Systems, Clinical/
27. decision mak*.mp. [mp=title, abstract, original title, name of substance word, subject heading word, keyword heading word, protocol supplementary concept word, rare disease supplementary concept word, unique identifier]
28. decision support.mp. [mp=title, abstract, original title, name of substance word, subject heading word, keyword heading word, protocol supplementary concept word, rare disease supplementary concept word, unique identifier]
29. 25 or 26 or 27 or 28
30. exp Qualitative Research/
31. exp Interview/
32. exp Focus Groups/
33. exp Anthropology, Cultural/
34. qualitative.mp. [mp=title, abstract, original title, name of substance word, subject heading word, keyword heading word, protocol supplementary concept word, rare disease supplementary concept word, unique identifier]
35. interview.mp. [mp=title, abstract, original title, name of substance word, subject heading word, keyword heading word, protocol supplementary concept word, rare disease supplementary concept word, unique identifier]
36. focus group*.mp. [mp=title, abstract, original title, name of substance word, subject heading word, keyword heading word, protocol supplementary concept word, rare disease supplementary concept word, unique identifier]
37. ethnogr*.mp. [mp=title, abstract, original title, name of substance word, subject heading word, keyword heading word, protocol supplementary concept word, rare disease supplementary concept word, unique identifier]
38. 30 or 31 or 32 or 33 or 34 or 35 or 36 or 37
39. 24 and 29 and 38
40. limit 39 to (english language and ("infant (1 to 23 months)" or "preschool child (2 to 5 years)" or "child (6 to 12 years)" or "adolescent (13 to 18 years)"))

Figure 1: Example Search Strategy

Screening of Studies

Searching and screening of studies is summarised in Figure 2. The title and abstracts of the papers retrieved from the searches were read by the lead author. Forty-six papers were retained following this initial screening and removal of duplicates. Forty-one of these studies were excluded based on the sample population, focus of the study, use of purely quantitative methods or because they were unable to obtain despite exhaustive efforts. Five studies met the inclusion criteria. The reference lists of the included studies were screened and one further eligible study was identified. On searching for this study, two other papers presenting different findings from this study were found and included. A further study by one of the authors (EP) also met the inclusion criteria and was included. A total of nine studies were included in the review.

Appraisal

There is some debate regarding the need to appraise qualitative research, however (Dixon-Woods et al., 2004) argue that the rationale for doing so is clear; to help the reviewer to appraise crucial aspects of the research, such as whether the methods and sample are appropriate to investigate the aim. Qualitative appraisal allows for the value of the research to be balanced against any methodological weaknesses (Edwards et al., 2000). A number of appraisal checklists exist; some of which have been evaluated and collated into one checklist by (Walsh and Downe, 2006). This was used in the synthesis to provide the authors with an overview of the limitations of individual studies and guide them in identifying the quality of included studies. Following appraisal, all studies were included within the synthesis.
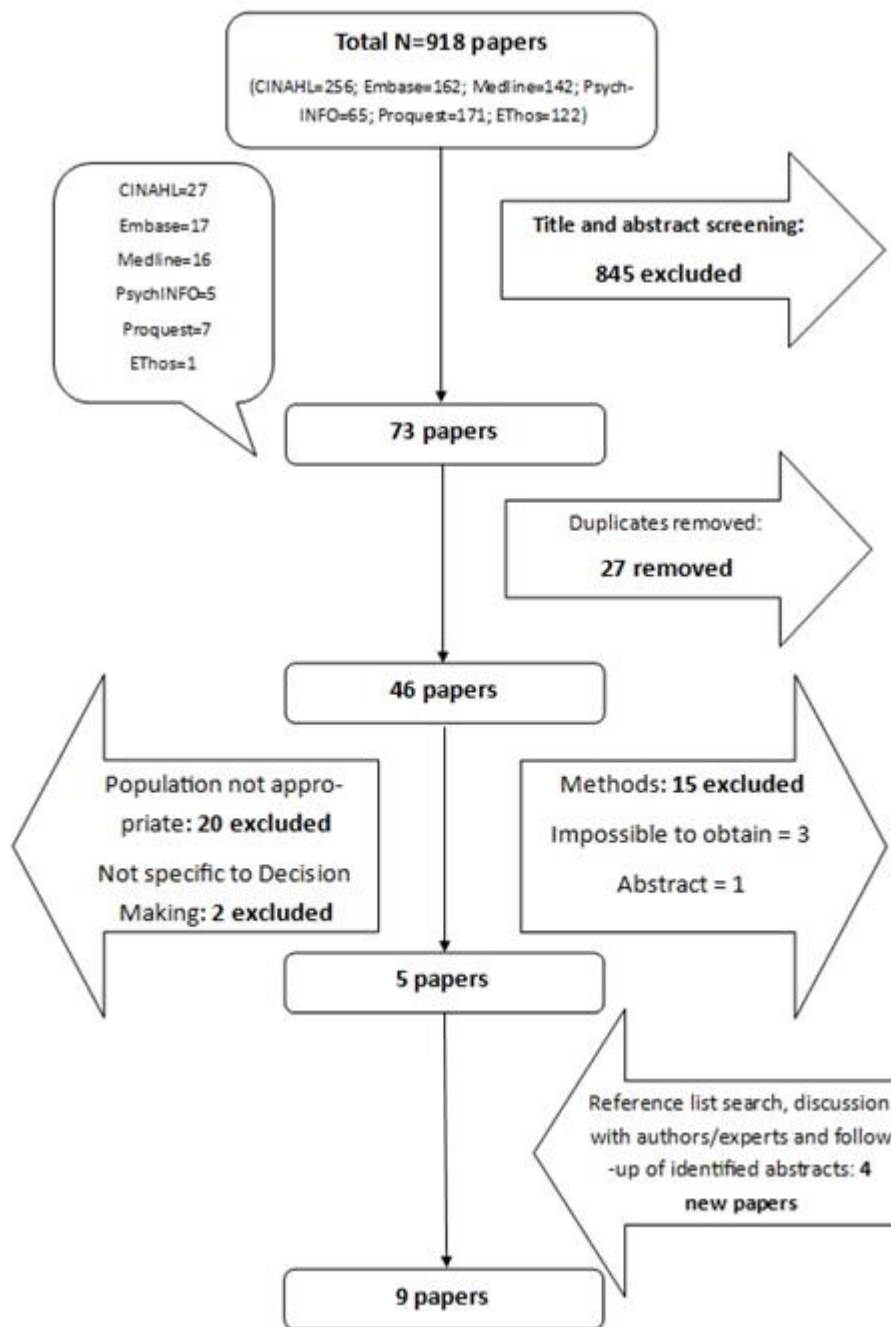
Figure 2: Detail of literature search

Description of papers

Included papers focussed on a range of stakeholders. The included studies collectively had a sample of 112 parents or guardians (henceforth referred to as parents for brevity), 35 children and 14 professionals. Three papers included only parents, two papers included only the child/young person and one paper included only health professionals. Only three

papers sought more than one perspective, two included parents and the child/young person and in one paper, cases were comprised of a mixture of the child/young person, the parent/carer and other family members. No studies investigated the perspectives of families and professionals simultaneously. Five papers included young people (up to the age of 37 years) and/or their parents. A summary of the papers is provided in Table 3.

Study, country	Purpose	Methodology and methods	Study Participants	Context	Main themes and sub-themes identified

Erby et al (2006), USA	Experiences of and attitudes to advance care planning for parents of DMD patients.	Exploratory, cross-sectional, semi-structured interviews.	Convenience sampling. 17 parents from 16 families of children and young people with DMD. Ages 8-37 yrs.	Recruited from Muscular Dystrophy groups.	1) Advance Care Planning (ACP) Communication
					2) Factors that influence ACP Communication:
					  a)Competing demands
					  b)Distancing from diagnosis
					  c)Absence of communication guidance
					  d)Stages of receptivity
					3) Navigating the unspeakable

Mitchell et al. (2011) UK	The realities of choice-making processes within families, exploring the respective roles and experiences of young people and their parents	Exploratory, longitudinal, semi-structured interviews.	Purposive sampling. 27 young people from 24 families and 44 parents from 33 families. Ages 13-22 yrs.	From 2 children's hospices.	Themed under the interview topics: 1) Young people's perspectives on making choices 2) Young people's experiences 3) Factors considered by parents regarding involving the young person 4) Moving on from full time education – experiences of parents 5) Factors and processes supporting satisfaction

Mitchell (2012) UK	Perspectives of disabled young people with degenerative conditions on making choices with parents and peers.	Exploratory, longitudinal, semi-structured interviews.	Purposive sampling. 27 young people from 24 families and 44 parents from 33 families. Ages 13-22 yrs.	From 2 children's hospices.	1) Parents' role in choice making
					  a)Nature of the choice
					  b)Which parent helped
					  c)Being listened to
					  d)Value of shared decision making
					  e)Parents' changing and evolving role
					2) Role of peers

Mitchell (2014), UK	To explore the perspective and experiences of disabled young people with degenerative conditions as they face significant medical interventions and engage in decision-making processes.	Exploratory, longitudinal, semi-structured interviews.	Purposive sampling. 10 young people. Ages 13-22 yrs.	From 2 children's hospices.	1) The decision
					  a)Why was the decision raised
					  b)Decisional Roles
					  c)How were the decisions perceived?
					2) The options
					3) Making the choice
					  a)Invasiveness of option
					  b)Uncertainty and risks
					  c)Quality of Life
					  d)Maintaining Health

					e)Retaining Independence f)Parents' wishes g)The actual decision 4) Reflection on choice
Mitchell and Dale (2015), UK	Experiences of PICU staff in advance care planning for children with LLI.	Exploratory, cross-sectional, semi-structured interviews.	Purposive sampling. 14 professionals (8 consultants and 6 senior nurses).	One PICU in Leeds.	1) Recognition of a LLI trajectory 2) ACP as a MDT structured process 3) Value of ACP 4) Inverse consequences of inadequate ACP
Nicholson (2012), UK	Decision processes in use of technological support in children with LLI.	Exploratory, cross-sectional, unstructured interviews.	Purposive sampling. 9 bereaved parents, 10 parents of children with limited/no decisional capacity, 6 parents of children with capacity and five young people. Ages 3-32 yrs.	From one children's hospice in England.	1) Decision Factors a)Weighing up the need for technological support b)Weighing up the potential outcomes 2) Decision Features a)Uncertainty b)The individual child c)Parents role as decision makers d)Beliefs and Values e)Gut instincts 3) Process Factors a)Background to decision b)Time c)Professional involvement d)Information for decision making e)The Healthcare system
Popejoy (2015) UK	To understand the 'lived experience' of parents throughout the process of making and revising end of life care decisions for their child.	Exploratory, cross-sectional, semi-structured interviews.	Purposive sampling. 3 bereaved parents from 3 families. Ages 3-15 yrs.	From one Children's Hospital in England.	1) Making decisions a)Timing b)Decision Makers c)Values 2) Revising and Implementing Plans a)Barriers and facilitators to implementation of plans b)Revising plans
Rapoport et al (2013), Canada	Explore experiences of bereaved parents regarding foregoing artificial nutrition and hydration (FANH) decisions for their child.	Exploratory, cross-sectional, semi-structured interviews.	Purposive sampling. 11 parents of 7 children from 6 families. Ages 1 month-15 yrs.	From one hospital. Identified from the Advanced Care Team.	1) Introduction of FANH to parents a)Being open to option in relation to perceived poor Quality of Life (QoL) b)Positive aspects of physician who

					introduced option
					2) Decision making process
					  a)Parental agreement
					  b)Importance of perceived family QoL
					  c)Childs perceived QoL
					  d)Importance of consistent message from professionals
					  e)Concerns about being judged

Wissow et al (2001), USA	Test acceptability and feasibility of undertaking advance directive development using a values history.	Exploratory, cross-sectional, semi-structured interviews	Convenience sample within one HIV service. At least 12 parents or guardians (reporting is unclear). 3 children participated. Ages 3-16 yrs.	Took place within normal clinic consultation, just somewhat structured interview to elicit values. Majority of decision makers were not biological parents.	1) Disabilities that would make life not worth living 2) Tolerance of Pain and Risk in Hope of Recovery 3) Past experience 4) Preferences for End of Life Care 5) Preserving life as long as possible 6) Spirituality

Table 3: Study characteristics

Synthesis

Data contained within a clearly defined results section were extracted for coding and all direct quotes were also extracted. Extracted data were coded in NVivo 10 using a data driven approach conducted by the lead author, and refined through an interactive process of constant comparison through discussion and written commentary to reach agreement between all authors. Codes were grouped into descriptive themes and then subsequently synthesised further into analytical themes (see Figure 3). The synthesis has provided a more comprehensive understanding of how decision making works in a wider context, considering the various factors that influence and constrain it, thus providing an understanding at the more analytical and conceptual level. Predominantly the individual studies had a much narrower focus in relation to decision making and the synthesis provided an opportunity to gain a broader overview of decision making. One study (Nicholson, 2012) identified many of the issues included within the synthesis and enabled the identification of these themes in the other studies, providing an opportunity for them to be unpicked and expanded on.
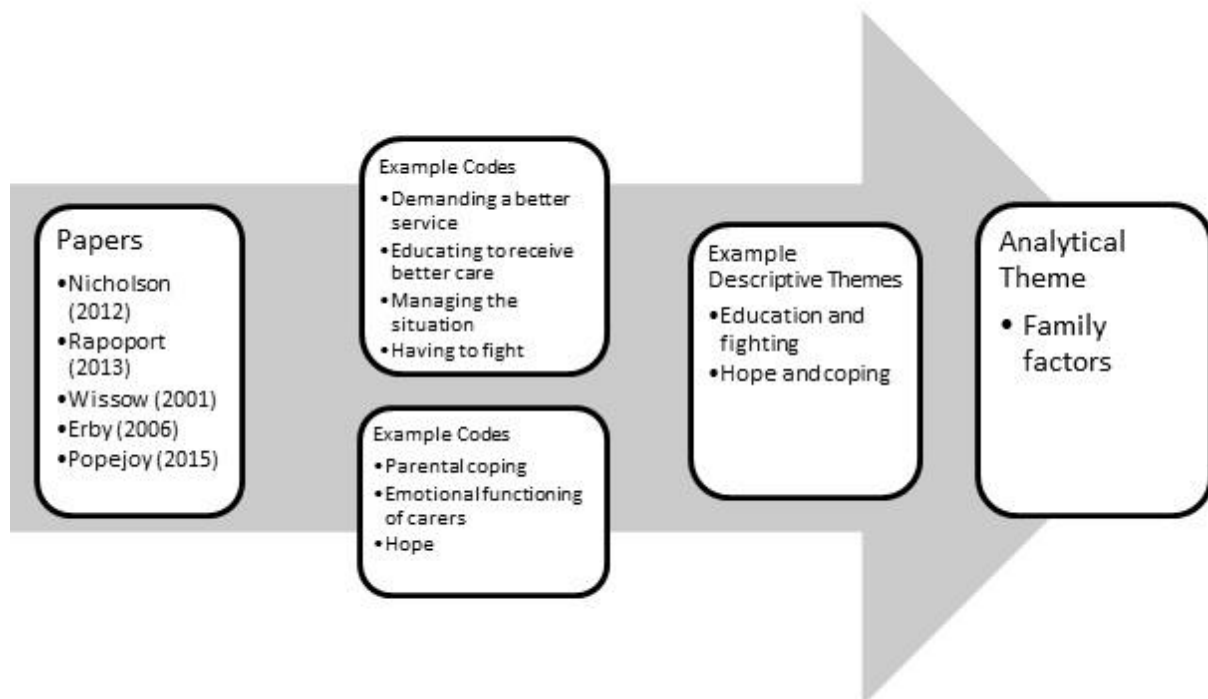

Figure 3: Example of the development of an analytical theme

Results

Nineteen descriptive themes were identified and four analytical themes developed from

these. The four analytical themes appeared to represent four different types of factors which influenced the decisions made by families and professionals: decisional factors, relational factors, family factors and system factors (Table 4). These four factors will be discussed below.

Theme	Description
Decisional Factors	Decisional factors were related to the specifics of the decision being made, including the consequences of a decision and the time available to make the decision. Underpinning descriptive themes: Timing of decisions, consequences, decisional considerations, difficulty in decision making, future planning.
Family Factors	Family factors impacted on decision making and depended on their background, culture and experience, awareness of the child's condition and their coping mechanisms. Underpinning descriptive themes: Coping mechanisms, awareness about illness, educating and fighting, family life.
Relational Factors	Relational factors involved individuals' perception of their involvement or choice in decision making, communication within the network of care and trust Underpinning descriptive themes: Gathering opinions, involvement in decisions, trust, agreement in decisions, communication and information exchange, support for decisions.
System Factors	System factors impacted on decision making through bureaucratic constraints and lack of access to support. Underpinning descriptive themes: Problems with the system, factors related to professionals, medicalisation, access to support.

Table 4: Description of Analytical Themes

Decisional Factors

Decisional factors encompassed a range of considerations specific to the decision being made. The extent to which families and children were given sufficient time to consider the decision and whether the decision could be predicted in advance, appeared to be important (Nicholson, 2012, Erby et al., 2006). Individuals held conflicting opinions about whether future possibilities ought to be introduced to them at an early stage or whether they should

be considered only when clinically necessary, as this impacted on different families' abilities to cope (Erby et al., 2006, Nicholson, 2012, Mitchell and Dale, 2015).

Decisions appeared to be considered in relation to previous decisions, interventions and the child's prognosis (Mitchell and Dale, 2015, Rapoport et al., 2013, Nicholson, 2012), rather than occurring as isolated incidents. The possible consequences of a decision, such as, aesthetic implications, the long or short term nature or reversibility of the decision were often considered (Nicholson, 2012, Mitchell, 2014), although individuals had differing perceptions about whether a decision was reversible. One young person identified having a gastrostomy as a decision with lifelong consequences (Mitchell, 2014), whereas other families considered this same decision as reversible (Nicholson, 2012). The child's level of comfort or quality of life had the biggest influence on the decision (Mitchell, 2014, Nicholson, 2012, Rapoport et al., 2013), but parents found the process of analysing the child's quality of life difficult (Rapoport et al., 2013). The difficulty in decision making was often made worse by emotional and physical exhaustion of parents' caregiving responsibilities (Nicholson, 2012, Popejoy, 2015).

Advance discussions and planning regarding limitation of treatment offered families time to consider what they want for their child (Mitchell and Dale, 2015), although it was acknowledged that such plans can and do change (Popejoy, 2015, Mitchell and Dale, 2015). Professionals viewed such discussions and plans as a communicative mechanism to ensure that care provided is in the best interests of the child and commensurate with the preferences of the family (Mitchell and Dale, 2015). The perspective of families regarding the purpose and benefit of such discussions was not identified within the reviewed studies.

Family Factors

Family factors impacted on the decision making process and the decision made. The child's character, views and their disease trajectory acted to influence the decisional process (Erby et al., 2006, Nicholson, 2012). Approaching specific stages of a known disease trajectory often precipitated decisional events. Nevertheless, a great deal of uncertainty existed, making planning for an uncertain future difficult (Erby et al., 2006). Family background was also important; those who considered themselves to have a good education and more life experience, both in terms of age and exposure to the health care system, felt that this helped them challenge professionals in decision making. There was discussion of the need to search for information and fight for the appropriate care for their child (Erby et al., 2006, Nicholson, 2012) which appeared to be more apparent where distrust in

professionals was expressed. Although family background was noted as important, it appeared that parental confidence in making decisions naturally increased over time (Nicholson, 2012).

The family's coping abilities and strategies are also influential on decision making, with some families feeling that they could not cope with certain decisional outcomes (Nicholson, 2012, Wissow et al., 2001). Discussing difficult decisions resulted in a period of decreased coping for some parents (Popejoy, 2015), which may lead to families resisting planning for the future in order to maintain their ability to cope with daily life (Erby et al., 2006). Other families used spirituality to help them cope (Wissow et al., 2001). Families identified that decision making is part of their life; requiring a delicate balance alongside managing the day to day care of their child and the practicalities of 'normal life' (Mitchell, 2014, Nicholson, 2012, Erby et al., 2006). Decisions were also considered in relation to their impact upon the wider family and other children (Rapoport et al., 2013).

Family awareness of the child's condition, prognosis and future possibilities was important in decision making. Families may not be aware of the illness trajectory (Wissow et al., 2001) or what intensive treatment involves or what the consequences would be (Mitchell and Dale, 2015). Families' developing awareness of the seriousness of their child's condition and the implications of their symptoms over time made decision making easier and allowed them to recognise the need for a particular course of action (Nicholson, 2012). Awareness was developed either by witnessing other children's illnesses or seeking advice from families who had previously made similar decisions (Rapoport et al., 2013, Popejoy, 2015, Mitchell, 2012, Mitchell, 2014, Wissow et al., 2001, Nicholson, 2012). However, some families tried to distance themselves from the diagnosis and associated community (Erby et al., 2006) so may not have had access to this type of support or developed awareness in this way. Professionals felt that they have an important role in facilitating the family's awareness and likely treatment outcomes by providing them with appropriate information (Mitchell and Dale, 2015).

Relational Factors

Families mostly felt involved in the decision making process (Nicholson, 2012, Popejoy, 2015), but sometimes felt that they were not listened to (Popejoy, 2015). Parents desired to involve their child in decisions (Erby et al., 2006, Nicholson, 2012, Wissow et al., 2001) and the child's feeling regarding the decision was frequently taken into consideration (Nicholson, 2012, Mitchell, 2014, Wissow et al., 2001). However, other parents were concerned about involving their child and protected them from this (Nicholson, 2012).

Some young people were or wanted to be actively involved in decision making (Nicholson, 2012, Mitchell, 2012, Mitchell, 2014, Wissow et al., 2001), whereas others did not wish to be involved at all (Mitchell, 2012, Mitchell, 2014) highlighting the importance of establishing individuals' preferences for involvement. When children reached the legal age to make their own decisions, parents sometimes felt excluded from decision making by professionals (Nicholson, 2012), although young people identified that they valued and desired their parents' input (Mitchell, 2012, Mitchell, 2014). Parents valued the input of professionals in decision making for their child (Nicholson, 2012). Involvement in decision making also related to the presence, or absence, of perceived choice and existed in situations where the child's condition dictated a particular action to be taken, for example, an unsafe swallow leading to termination of oral feeding (Nicholson, 2012). (Nicholson, 2012) identified that even in situations with little perceived choice, the process of agreeing to a treatment was important for families as it required them to understand the risks and benefits involved and come to their own conclusion that the treatment is necessary.

Communication and information exchange played an important role in decision making. Some families expressed problems in accessing information due to the rarity of their child's condition or lack of openness by professionals (Nicholson, 2012), whereas other families received open and honest information about the available options (Rapoport et al., 2013). Professionals expressed concern that often communication with families regarding the implications of intensive care treatment for their child is poor (Mitchell and Dale, 2015). Quality of communication between professionals and the child varied (Nicholson, 2012, Mitchell, 2014, Erby et al., 2006) and families appreciated when their knowledge of their child was welcomed and valued by professionals (Nicholson, 2012), but this did not always occur (Popejoy, 2015, Nicholson, 2012).

Trust within the family-professional relationship was important in facilitating candid conversations with the child and family (Erby et al., 2006) and enabled professionals in certain circumstances to take the burden of decision making away from the parents (Popejoy, 2015). Many instances were identified where a lack of trust was demonstrated and occurred when professionals did not listen to parental concerns or had previously made errors (Nicholson, 2012). In the absence of trust, parents suspected a hidden agenda when treatment decisions were discussed (Nicholson, 2012), which may have impacted on their ability to make an informed decision. Agreement between various stakeholders in the decision also appeared to be important; both agreement within the family, between the family and professionals and within the healthcare team (Rapoport et al., 2013, Nicholson, 2012, Mitchell and Dale, 2015). Professionals identified that it can be difficult to achieve consensus regarding the appropriate treatment for each child and the

difficulty in ensuring that discussions have been had with families prior to an acute deterioration (Mitchell and Dale, 2015).

System Factors

System factors were identified as important and came through strongly in both (Mitchell and Dale, 2015) and (Nicholson, 2012) and referred predominantly to health care systems, but also occasionally to social and educational systems. Some families felt that there were too many professionals involved in the care of their child, which made continuity and effective communication difficult (Nicholson, 2012). Rules and procedures often made decision making or accessing care difficult and prevented an individualised service (Nicholson, 2012). Additionally families encountered errors within the healthcare setting, such as medication errors, surgical complications or inability of professionals to notice the subtle signs of a deterioration in their child's condition; often families made the decision to stay with their child continually during a hospital admission, due to lack of confidence in staff (Nicholson, 2012). Additionally, professionals identified that likely deteriorations in the child's condition are often not anticipated or planned for appropriately and children are subsequently given inappropriate treatment, whereas better planning may have resulted in more appropriate outcomes (Mitchell and Dale, 2015).

Another issue related to the healthcare system was the difficulty in accessing advice or support. Families identified the difficulty in obtaining support and advice from their main doctor (Nicholson, 2012) which may have left them to make decisions alone. Other families expressed that their doctor made time for families when required to discuss any issues (Nicholson, 2012). Families articulated that there is a difference between 'good' and 'bad' professionals who will either help them to navigate the system and overcome the rules, or who rigidly apply the rules (Nicholson, 2012). When families found a 'good' professional, they knew that they could rely on these people to help them make decisions and access the treatment their child needed (Nicholson, 2012). Professionals themselves acknowledged the problems of the system where care is often over medicalised and professionals become emotionally exhausted through providing care and dealing with death (Mitchell and Dale, 2015).

Discussion

This thematic synthesis has identified numerous influential factors to decision making for children with life-limiting conditions. These were broadly categorised as: decisional factors, family factors, relational factors and system factors. Options were considered in relation to previous decisions made, the potential consequences and the reversibility of the option. Specific factors relating to the child and family, also impacted on the decision, for example, the disease trajectory, coping strategies and awareness regarding the condition. Trust

between the family and professionals played a large part in decision making, as did information provision. The stakeholders involved in decision making varied between different families. Finally, system factors, for example, rules and procedures or access to support, could either facilitate or hinder the making or implementing of a decision. Decision making for this population involves the complex interplay of many factors. Although some aspects of the decision making process may be different for children with life-limiting conditions, it is likely that there is some overlap for children with complex illnesses and disabilities. The identified factors will be considered in relation to some of the literature on children with life-threatening or chronic illnesses in order to identify any similarities and differences.

Within the wider chronic or life-threatening illness literature, decisional factors have also been identified as important in decision making. Preference for the timing of the introduction of discussions appears to be individual for each family (Gibson, 2001, Jackson et al., 2008, Dellon et al., 2010, Edwards et al., 2012, Gotz et al., 1997, Noyes et al., 2013) and may impact on perceived choice in the decision (Sharman et al., 2005). Prognostic uncertainty also influences decision making, making it more difficult (De Graves and Aranda, 2005, Kirschen and Walter, 2015).

Family factors were noted to influence decision making and these included experience and awareness of the disease trajectory. Although not specifically identified within the reviewed papers, it is possible that the multiple phases of the child's illness and previous experience of the child's ill health link into these and impact on decision making. Changes in the child's disease trajectory are viewed as significant by families (Wood et al., 2010) and influences the way they view the disease (Bluebond-Langner et al., 2016). It follows that this may factor into the decisions that families make, but this requires further investigation.

Other family factors have also been noted also in the wider literature, with family characteristics, such as hope and the child's QoL, influencing decision making (Maurer et al., 2010, Higgins and Kayser-Jones, 1996, Tomlinson et al., 2011, Hinds et al., 2000, Lan et al., 2007, Tomlinson et al., 2006). The role of these characteristics however, appear to be different between the thematic synthesis and wider literature. For children with cancer, families pursue cure-directed therapy even when the chance of cure is minimal, as they try to extend the child's time and not give up hope (Bluebond-Langner et al., 2007). Although hope was identified in the thematic synthesis, it did not appear to impact on decision making, rather assisted families to cope with life and the child's illness on a day

to day basis. Anticipation of the child's future in light of a life-limiting diagnosis appeared to be different than for those with a potentially curable illness.

The wider body of literature, including children with life-threatening and chronic illnesses has identified the need for families to seek information on their own in order to make informed decisions (Watson et al., 2002, Bluebond-Langner et al., 2007) and exert pressure on health professionals to ensure their preferred course of action is taken (Lindblad et al., 2005, Russell, 2003). This seems to reflect the descriptive theme within the synthesis of 'Educating and Fighting' and may reflect the need for those in regular contact with the health care system to have their individual needs recognised.

Additionally, relationships have been identified in the wider literature as important in decision making. Primarily families feel that decision making is their responsibility (Carroll et al., 2012, Sullivan et al., 2014), but despite this, families value the input and guidance provided by professionals (Rodriguez and King, 2014). The thematic synthesis suggested that families want to be involved in decision making, but did not identify who they considered held the ultimate responsibility for this. The inclusion of children in decision making has also been discussed, although the majority of this research has been conducted with children and young people who are healthy, have HIV or cancer (Berger, 2013). From the limited research in this area, it appears that there is a consensus that young people have the right to be involved in decision making and future planning discussions (Pousset et al., 2009, Talati et al., 2010). The literature suggests that such discussions are safe to have and increase decisional congruence between young people and their families (Lyon et al., 2010a, Lyon et al., 2010b, Lyon et al., 2009a, Lyon et al., 2009b, Lyon et al., 2013). These findings may not be directly transferrable to children with life-limiting conditions, as many have some degree of cognitive impairment and may need additional support to participate in such discussions. Further research is needed to identify what role children and young people with life-limiting conditions have in making decisions about their care and treatment and the impact of their age and prognosis on their involvement.

The lack of acknowledgement by professionals of families' specific expertise regarding their child has been reported in relation to paediatric chronic illness (Lindblad et al., 2005, Watson et al., 2002, Graungaard and Skov, 2007). This may impact on other relational issues such as trust, which has also been highlighted as an issue in the family-professional relationship for children with chronic illnesses (Graungaard and Skov, 2007, Lindblad et al., 2005, Russell, 2003). However, as would be expected there is variability in the quality of professional relationships and communication, with caring and trustworthy professionals

also being acknowledged (McMaster et al., 2004, Nuutila and Salanterä, 2006, Moore and Kordick, 2006, Konrad, 2008). Trust and acknowledgement of the family's expertise was identified within this synthesis as a descriptive theme relating to difficulties in decision making. The specific impact, in relation to the process of decision making, of trust and acknowledgement of parental expertise needs further investigation.

System factors have been highlighted within the chronic or life-threatening illness literature in relation to incidents of medical errors (Sacchetti et al., 2000, Srivastava et al., 2005, National Reporting and Learning Service, 2009, Chua et al., 2010), rather than the impact that these factors have on decision making. This synthesis has identified that errors in hospital and problems accessing support and advice, impact on decision making. Further research is needed to identify how system factors are considered within decision making.

As can be seen from this discussion, the analytical themes identified within this synthesis have some commonalities with the wider literature on chronic and life-threatening conditions, however the manifestation of these themes appear to be different for those with chronic, life-limiting or life-threatening conditions. This synthesis has revealed the individuality of family preferences for engaging in decision making and future planning, but the included studies provide no indication of how professionals can best discuss and identify these or ensure that these are accommodated. Further research investigating how professionals might identify and meet family preferences for involvement is required. Additionally this synthesis has shown that future planning has not been a central focus within the literature so far. Included studies focussed only on ACP, suggesting that the concept of future planning, more broadly, requires further investigation.

The themes resulting from this thematic synthesis are similar to those identified by (Nicholson, 2012), which is unsurprising given its weighting in terms of data for consideration in the synthesis. However, although (Nicholson, 2012) recognises the individual role of professionals and families, this is presented from the family's perspective only and there is little discussion of the interaction between them and its impact on decision making. The model of decision making proposed by (Nicholson, 2012) suggests that process and decisional factors are separate systems which rarely interact. This synthesis however seems to suggest that professionals may act as a bridge between these two systems.

Limitations

As with any literature synthesis, the results are limited by the data reported in the included studies, which are necessarily a 'snapshot' of the data collected. However, the inclusion of individual study authors' findings in the synthesis (Thomas and Harden, 2008) aimed to mitigate this limitation as the authors will have had an overview of the whole data set before analysing it into themes.

This synthesis aimed to provide insight into the process of decision making for children with life-limiting conditions. Papers investigating decision making for children with life-threatening conditions were excluded, although as highlighted in the discussion there may be some similarities and differences between these groups. As the wider literature enabling this comparison was not identified through a systematic search, it may be that there are more similarities between the groups and that excluding the studies investigating children with life-threatening conditions has prevented deeper insights into the process of decision making from being revealed.

Finally, the lack of multiple perspective research in this area and the dominance of the family or parent view in the included studies mean that little is known about the relational aspects of decision making. Additionally, the included studies were predominantly small and cross-sectional in nature and although two of the papers presented data from one longitudinal study, they only presented data from one time point, meaning that the dynamic process of decision making and future planning could not be analysed. The evidence base in this area is limited and further research is needed.

Conclusion

This is the first synthesis of the qualitative literature regarding decision making and future planning for children with life-limiting conditions. The results indicate that there are a number of important factors to consider when caring for families of children with life-limiting illness and specifically when initiating conversations about decision making and future planning: decision factors, family factors, relational factors and system factors. The synthesis identified that decision making and future planning needs to be individualised for the family and their circumstances. Clinicians need to be aware of how aspects related to the specific decision and the family's background may impact on their choice, and assist them in information seeking. Families were satisfied when they were involved in discussions and their expertise acknowledged, therefore professionals should aim to communicate effectively with families to identify their preferences for involvement and initiation of discussions. Relational factors were identified as important, thus highlighting

the need for professionals to consider how to improve and maintain their relationships, thereby supporting information sharing and the development of trust. However, more research is needed to provide definitive recommendations to professionals regarding how to individualise care regarding decision making. Further research should also investigate the extent to which each individual is involved in the process and the relational and longitudinal nature of decision making.

Quotes from included papers to illustrate themes

Theme	Sub-theme	Exemplary quotes
Decisional Factors	Timing of decisions	"Thinking too much, I think that gets you into a lot of trouble. That is what I kind of adopted the philosophy of. You kind of deal with it one day at a time and you deal with whatever needs to be dealt with at the time. Because I think if you constantly ponder on the future, I don't think you would ever be able to function on a daily basis. I wouldn't be able to function on a daily basis." (Erby et al., 2006 - Parent) Not all were first-time decisions. For half, the decision had been previously considered and rejected or postponed, leading it to become an on-going choice to be revisited when (medical ⁄health) circumstances demanded or the young person initiated revisiting the decision. (Mitchell 2011 – Results Section) There a distinction between the overall decision process, which can last many years as a child's symptoms worsen and parents continually assess their child's needs, and the actual process of deciding, the latter of which is defined as the time during which families weigh up the appropriateness of technological support, and acquire the information they need to make an informed decision. For some families, the duration for these different aspects of the decision process is the same, and this was certainly the case for families who had not expected a recommendation for technological support to be made. (Nicholson, 2012 – Results Section)
	Consequences	"When he used to have that tube in his nose and he was always pulling it out, and it was always getting sore and we were always having to put it back in his stomach. It was horrible. It was just horrible you know, having to go through that every time it came out or dislodged itself. So you know, in that sense, it made having a decision, a PEG or a button, it made it easier really in that sense. Because you didn't want to see that all the time." (Nicholson, 2012 - Parent) A minority of parents 'framed' the decision in terms of finding something local to occupy the young person during the day. However most drew on notions of 'normality', talking about their son/daughter's right as a young adult to move on in life, to make their own choices and achieve maximum independence within the challenges set by their physical impairments and (where relevant) learning difficulties (Mitchell et al., 2011 – Results section)
	Decisional considerations	The young people engaged in a complex process of trade-offs, considering both the costs/risks and benefits to their health of accepting a medical intervention, alongside broader social and emotional quality of life factors such as the importance of 'normality' and preserving nonmedicalised aspects of their life. Choosing when to accept an intervention often involved balancing these needs or aspirations with medical necessity, sometimes even life-preservation. (Mitchell, 2014 – Results section)

		"[H]e's never going to see, hear, walk . . . you feel you're as bottom as you can go . . . how do you as an individual ultimately interpret that? Is that what you really want? Or is that good for, for [child]? How does that play out in your life plan and what does it mean for everybody else, including [daughter], you know? Our other daughter and ourselves as well, still as a family, how's all this gonna work if you go down 'route A' as opposed to 'route B'?" (Rapoport et al., 2013 - Parent)
	Difficulty in decision making	Multiple barriers to successfully managing the process were identified, including time constraints, conflicting clinical demands and lack of formal training in communication skills. In the PICU setting, a lack of established rapport with the family before having to raise the issue of end-of-life care for the first time during acute situations, including resuscitation, was identified as causing particular difficulty (Mitchel and Dale, 2015 – Results section) "And we're exhausted at that point, absolutely at the end of our tether, you know, and then making these decisions when you're in that state. In this totally, mentally, physically exhausted state, and that's not taken into consideration all the time." (Nicholson, 2012 - Parent)
	Future planning	"if they make an ACP they may still change their mind right at the very end, um … But at least they will have had the opportunity to sit down and seriously think about what they want for their child." (Mitchell and Dale, 2015 - Nurse) There was some evidence in the data that acknowledgement is more likely for children and young people with conditions where it is expected there will be feeding or respiratory problems, especially when compared to children with undiagnosed or very complex conditions. For these families, a child's swallowing and respiratory function is sometimes monitored for years, and the decision was described by some families as a planned one, gradually introduced to families at different time points before being initiated at the appropriate time. In fact some young people recall always knowing they would need ventilation and therefore accepting it as inevitable, and do not always identify having made a decision about ventilation per se, but rather about the timing, type, and location of initiation (Nicholson , 2012 – Results Section)
Relational Factors	Gathering opinions	"Our end decision would have still be mine. But it helped having somebody else's input, other people's decisions coming in as well.....You know their points of view came in as well and I thought yes. You know sometimes when you're in that situation you can't think, you can't always think straight really can you, sort of think what's best, and so having other people's decisions made it slightly easier for me ." (Nicholson, 2012 - Parent) I mean I do listen to mum a fair bit, I mean it's my decision in the end but I do listen to mum a lot .. . cos she's very clever, I don't know it just, well she's my mum and she knows best ... yeah, I trust my mum's opinion. (Mitchell, 2012 – Young Person)

	Involvement in decisions	Within the discussion of young people's developing capacity, it is clear that parents play a central role in facilitating their child's participation in decisions that affect them. However, like children, parents embark on their own journey as decision makers, and this in some ways resembles the journey children make in that parents also develop capacity for decision-making over time, and the parents in this study provide remarkably similar accounts of moving from being dependent on the healthcare professionals involved in their child's life to assuming control of the decisions that need to be made for their child. (Nicholson, 2012 – Results section) "Well now as I'm getting a lot older they'll [family] listen to me but like when I were younger, five or six like, me mum had to make decisions for me and so did the hospital but now, like when I got to, like 10, that's when me mum and our [brother] started saying like 'no, do you want to do this?'" (Mitchell, 2012 – Young person)
	Trust and confidence in staff	"The only one time we didn't stay, she just, she lost a day of her life because somebody messed up with drugs…..So she's never been left after that. I mean cock ups have happened since, with the drugs, I mean they're strong drugs….that's why whenever she goes in now, we always say "we'll medicate Emily"." (Nicholson, 2012 - Parent) "[S]he had that casual and confident way about her that just makes you feel that she knows what she's talking about. You have faith in her, and, it just makes it all work." (Rapoport et al., 2013 - Parent)
	Agreement in decisions	"At some point, once you realise that you have a better idea of what's wrong with your child, and how serious it is, you can say "well actually, we respect your opinion but on this occasion…."…..we grew to trust ourselves to make the right decisions for Robert." (Nicholson, 2012 - Parent) "before you can convince any parents, you have to convince the other specialties. You have to bring them on board. If they're not on board, you have no chance, or your chances with the family are much much less." (Mitchell and Dale, 2015 - Doctor)
	Communication and Information exchange	"It was quite hard hitting, it was quite honest questions, that actually you know it might be that, [now referring to the consultant] "I've got to be honest with you Lee. I might actually put you under anaesthetic and actually, we might not be able to do it because, can you see from these x-rays, you know, some of your bits aren't in all the right places"." (Nicholson, 2012 - Parent) Health care professionals who were helpful to parents acknowledged the child's perceived poor QoL and feeding difficulties and explained why FANH (Foregoing Artificial Nutrition and Hydration) was a reasonable option. Parents appreciated that the option was discussed in a sensitive, nonjudgmental, and supportive way. They also appreciated being given enough time to reach their decision; within hours to a few days of being presented with the option all parents concluded that FANH was best for their child. (Rapoport et al., 2013 – Results section)

	Information and Support	"The person who's on the ventilator understands because they're on it. They know how they feel." (Nicholson, 2012 - Parent) Written information about the procedure, or supplementary information found on the internet or provided by third party organisations such as condition specific charities is useful but it does not make up for the invaluable information and opinion provided during discussions with healthcare professionals, and with other families themselves (Nicholson, 2012 – Results section) Young people without LCIs felt that peers with similar conditions could be a source of 'inside information' and/or emotional support. 'Inside information' was regarded as 'lived' or experiential information from a person who had previously faced a similar choice. This was valued as it was a type of information which the young people felt professionals or parents could not provide. (Mitchell, 2012 – Results section)
Family factors	Coping	"sometimes I think I talk about things and I start to cry, and I don't know why….I get so upset. [Child's name] ain't going to get better. I don't know what to do, I'm doing the best I can, but I start to talk about it and my chest gets so tight. I don't know what I can do to help him, and the next thing I know I'll be crying." (Wissow et al., 2001 – Child's Guardian) When hope was not accessible to parents, some expressed a tendency to get from day to day by avoiding the reality of muscular dystrophy to some extent. (Erby et al., 2006 – Results section)
	Awareness	"Dad: "We didn't realize. We thought, oh it's just a bit of food. It's got in with her snot basically. We were thinking things like that. And it were getting worse wasn't it? Mum: "Yeah but because she were eating it didn't matter."" (Nicholson, 2012 - Parent) Over time, however, Sebastian's parents noticed a change in Sebastian's breathing overnight, and they were unsure of the reasons for this. This development triggered Sebastian's parents to seek medical advice, and this point can be seen as the start of the decision process that led to overnight ventilation. (Nicholson, 2012 – Results section)
	Educating and fighting	"So I took them all kinds of stuff. All kinds of literature and stuff, and I kind of just threw it on the table at this meeting and said, 'It's kind of like your job to just read it.' " (Erby et al., 2006 - Parent) "You have to have all the information to make the right choices, especially if it's a child, for your child. So you have to fight. Don't just sit back and take their word for it. You have to fight." (Nicholson, 2012 - Parent)
	Family life	"A lot goes on behind the scenes, it's like a fabulous film but nobody knows what goes on to make it … and it's a lot of work, a lot of stress and, in-between all that, people are trying to live normal lives aren't they, you know, do the

		supermarket shopping and take the kids to the dentist … all the mundane things you do as a parent … I do a lot of juggling, I keep a lot of balls in the air." (Mitchell et al., 2011 - Parent) "You look at trees differently. You embrace life in a different way. We have done more traveling because we feel now that [our son] has to experience everything before he runs out of time to experience stuff, you know? We'll have dessert for dinner. We kiss the monkeys. You're not in pain. Everyone else is so serious about ... we just let our children be children and we live a wonderful life because of [our son]." (Erby et al., 2006 - Parent)
System Factors	Problems with the system	As well as the loss of key resources for decision-making, families talk about the separation of services due to the different infrastructure making up adult health care. Some young people will have several new consultants, none of whom take overall responsibility for their care, and none of whom specialise in their condition, instead each having an organ or function which they accept responsibility for. In some cases, these professionals can be based at different hospitals therefore further fragmenting the care young adults receive. For families who are able to maintain a relationship with one of the professionals they have come to trust over many years, this resource is highly valued, and can become the primary contact for on-going problems their adult child may now experience. (Nicholson, 2012 – Results section) "Now it may have been August time but it required several people to liaise in order to make a decision about providing oxygen but they were on holiday at different times....and every night Sebastian had this problem so it's incredibly cruel and reflects this indifferent system of rule following.....Anyway, eventually they all came back, had a consultation. And it was agreed that yes Sebastian certainly did need oxygen." (Nicholson, 2012 – Results section) "These organisations are like super tankers…..They've got a momentum of their own and they don't, they're inflexible. They can't cater for special circumstance. They're sort of like sausage machines. So children like Sebastian, they can't cope with." (Nicholson, 2012 - Parent)
	Professionals	"when I was a registrar it was easier for me because I had to just sit and have a debrief with my … consultant, as I love to cry. But now, I have to be this brave person and it's very … very difficult." (Mitchell and Dale, 2015 - Doctor) "Anybody who says the NHS works hasn't experienced it properly….I reckon 80% of the staff who work in the NHS aren't that interested….They turn up, do their job, go home, forget about it. The other 20% actually care. But you've got to find them." (Nicholson, 2012 - Parent) "By the time they've [nurses on the ward] done 12 hour shifts. If it's full on, by time you've got to 10 hours they're absolutely zonked. It's too long…..Three days….Three 12 hour shifts. 36 hours that's it. Get it all done in three days…..but then if you got a good nurse, where you were confident with, and you thought, bloody hell she's good." (Nicholson, 2012 - Parent)

	Medicalisation	"So I think there are many examples in my head and in my career where we have over-medicalised the end-of-life and families have clung on to the medical management rather than the holistic palliative care management of children, such that they feel that they're failing their child if they don't ask for everything." (Mitchell and Dale, 2015 - Doctor)
	Access to support	"It may well have been that I phoned the consultant, I was told by the secretary of one consultant, "consultants don't talk to the patients. You know, you can't ring up and have an informal conversation". So when you're meeting barriers like that it's difficult. We had a heck of a struggle to rectify, to overcome this problem." (Nicholson, 2012 - Parent) "At the end of the meeting he pushed his card across and he said, and he turned it over and on the back was written his home telephone number. And he said "you've a difficult time, any problems ring me at home." Now that obviously is the sort of person you want. Not some-one who thinks, I am, you know, mini god." (Nicholson, 2012 - Parent)

- **Key messages**

  - Four factors impact on decision making: decision factors, family factors, relational factors and system factors.

  - Professionals should identify family preferences for decisional involvement and individualise communication and engagement to meet their preferences.

  - Further research is needed regarding the contextual, relational and temporal aspects of decision making for children with life-limiting conditions.

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