Title page

Title: Concurrent transitions: A systematic review into the life transitions of adolescents and young adults with life limiting conditions: impact on the person and their significant others

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Abstract

Purpose: A systematic review was conducted to appraise and classify evidence related to the life transitions of adolescents and young adults with life limiting conditions.

Methods: Databases searched were MEDLINE, CINAHL, PsycINFO, CancerLit, and AMED. Methodological quality was assessed using an established tool, and final included articles were rated as moderate to high quality. Articles were then assessed based on the insight that they provided into life transitions for adolescents and young adults.

Results: Eighteen studies were included in the final review, with two major life transitions identified as pertinent: “illness transition” and “developmental transition”. These concurrent transitions were found to be relevant to adolescents and young adults with life limiting conditions, generating complex needs. Sub-themes within the transitions were also identified, enhancing understanding into them. Furthermore, the illness transition was found to impact on significant others, namely family members, requiring them to make adaptations and had physical, mental and emotional health implications.

Conclusions: Future research is needed to focus on adolescent and young adult perspectives to bring further insight into these key transitions, since such

perspectives are currently underrepresented. Attention to the impact of the illness on the whole family would be useful to expand findings from this review.

Keywords: Adolescents, young adults, life limiting conditions, palliative care, end of life care, life transitions, systematic review

Implications

This review found that “illness transition” and “developmental transition” were key concurrent transitions for adolescents and young adults with life limiting conditions. Illness transition had a number of impacts upon significant others such as family members. Further research is required to expand understanding and evidence to improve care.

Introduction

Increasing awareness and emphasis on the needs of adolescents and young adults with life limiting conditions is emerging(Marsh et al., 2011). With advances in medical and nursing care, many children are living longer with conditions that were previously unique to childhood (Doug et al., 2011, Marie Curie Cancer Care, 2012). The care of these adolescents and young adults can often be more complex due to the fact that they are growing and developing within the continuing presence of their illness and changing prognosis. This may mean that help, support and interventions are required from a health, social and educational professionals and services.

Adolescents and young adults with life limiting conditions are likely to be undergoing several life transitions(Jindal-Snape and Miller, 2010) which largely revolve around developmental, educational, employment and relationship issues. This review explores how life transitions are experienced by adolescents and young adults, in the context of them having a life limiting condition and, therefore, how these transitions inform their psychosocial needs. It is important that this is understood by health and social care services, and other relevant professionals, in order to provide optimal support to both the young adults and their families. Adolescents and young adults with illness still desire to achieve independence, attain educational achievements, live independently and have a social life (Marie Curie Cancer Care, 2012) (Muscular Dystrophy Campaign, 2010). Moreover worries for adolescents and young adults with serious illnesses may be focused on adult-related milestones, such as not fulfilling one's potential, as opposed to health issues (Muscular Dystrophy Campaign, 2010).

This review also provides insight into effects of life transitions on significant others, i.e. key people present in adolescents' and young adults' lives such as loved ones (family, friends), care professionals and teachers. This helps address the difficulties that loved ones may face. One report (Muscular Dystrophy Campaign, 2010) highlighted that some young adults' worries may focus on how diagnosis will affect their family members rather than themselves. The little research that has been done suggests that families' needs often go unmet by health services (Aruda et al., 2011). Therefore, understanding into impact on significant others, such as family members, is needed to clarify their needs and find ways of addressing these.

Previous reviews

Previous reviews on research related to adolescents and young adults with illnesses have focused on transitions in the sense of transitioning from child to adult services (Betz et al., 2013, Fegran et al., 2014, Jordan et al., 2013). For instance, one review (Bloom et al., 2012) explored the health care transition for youth with special health care needs. The findings suggest that adolescents and young adults with special care needs experience smooth transition to adulthood when they have less complex and milder conditions. Their largely quantitative systematic review also found that little has been reported regarding interventions that may help a smoother transition process to adult services. Likewise, Doug et al. (Doug et al., 2011) also review the evidence around the transition process for adolescents and young adults with palliative care needs, focusing on the move to adult services. They found that that transition programs to aid the process to move to adult services were not standardised or grounded in good quality empirical research and evidence base. Moreover, transition models were largely specific to particular conditions such as Cystic Fibrosis, as opposed to being appropriate for more general palliative care needs. Nevertheless, Doug et al.'s review does acknowledge the importance of family members in supporting significant transitions, yet, does not focus on the needs of family members.

Our review departs from previous work by taking a more focused lens towards the movement to adulthood as opposed to movement to adult services. We recognise that the developmental transition is a key transition for this age group, revolving

around aspects of personal development. Past work has briefly addressed developmental aspects within the broader “transition to adulthood” theme (Bloom et al 2012). In addition, our review also acknowledges and examines the importance of the illness transition as existing concurrently with the complexities of becoming a young adult. It explores the experiences of adolescents and young adults with life limiting conditions and draws upon largely qualitative data to explore in detail the impact of transitions on the young person and their significant others.

Aims

The overall aim was to understand the way that adolescents and young adults with life limiting conditions experience life transitions and to explore the impact on them and their significant others.

Research questions

1. What life transitions do adolescents and young adults experience in the context of having a life limiting condition?
2. What is the effect of their life transitions on their significant others?

Terminology

For this review, “adolescents” depict those aged between 10-19 years of age, as advised by the World Health Organisation (WHO) (World Health Organisation); Young adults depict those aged between 20-25 as guided by other studies looking at this group (Lau] et al., 2014, van Staa and Sattoe, 2014).

Our use of “life limiting condition” is informed by the Association for Children’s Palliative Care (ACT) (Association for Children's Palliative Care (ACT), 2009), in understanding this to be any condition known to shorten people’s lives. We conceptualise transition as an on-going process of moving from one context and set of interpersonal relationships to another (Jindal-Snape and Miller, 2010).

Methods

We conducted a systematic review within the time period 2000-2015 to identify key articles describing studies which would enhance understanding into the focus of the research questions. Once retrieved, these studies were appraised and assessed based on their ability to inform our research questions.

Inclusion criteria

Inclusion and exclusion criteria were drawn up to establish identification of focus, purpose and variables of interest. Please see Table 1 for details of these criteria.

[Insert Table 1 here]

Search strategy and article selection.

Medical Subject Headings (MeSH) key words were identified and were developed into a search strategy along with associated free text terms. This strategy was used

to perform a systematic search of selected databases (MEDLINE, CINAHL, PsycINFO, CancerLit, AMED). Please see Table 2 for the search strategy as developed for MEDLINE. Articles were selected based on their ability to meet our inclusion criteria. Figure 1 provides a “Preferred Reporting Items for Systematic Reviews and Meta-Analyses” (PRISMA) flow diagram, which captures the phases of the systematic review undertaken to reach the final number of included articles. Guidance for producing the PRISMA were taken from the PRISMA website.

[Insert Table 2 here]

[*Insert Figure 1 here]*

Assessment of methodological quality

As recommended by the National Collaborating Centre for Methods and Tools (Ciliska et al., 2008) the Critical Appraisal Skills Program tools (CASP) were used to consider methodological quality. Each of the ten CASP criteria was scored 0, 1 or 2, with a score of 0 meaning not or inadequately addressed; a score of 1 meaning the criterion was partially addressed, and a score of 2 meaning the criterion was fully addressed. Papers with a total score of between 15 and 20 were considered to be ‘high’ quality, ‘moderate’ papers scored between 8 and 14, and ‘low’ quality papers were those scoring between 0 and 7. Papers were assessed by two reviewers independently, who then agreed the final rating. Final included papers were rated as moderate to high quality.

Results

A total of 18 studies were included in the final review. Please see Table 3 for an overview of the included studies, including participants, ages of young person with the life limiting condition, nature of the life limiting condition and themes.

[*Insert Table 3 here*]

The two key life transitions for adolescents and young adults that studies focused on were “Illness transition” and “developmental transition”. Illness transition revolved around the way in which the young person adapted to having an illness or illness progression and the activities and responses around this. This led to sub-themes of *illness trajectory;* and *coping strategies.* The findings suggested that “developmental transition” encompassed several aspects around personal development as demonstrated in the sub-themes: employment*/education/training*; *future planning; relationships; independence*. The two major transitions and their related sub-themes are conveyed in Figure 2.

When exploring the impact of illness transition on significant others, namely family members, six categories were identified which helped summarise findings: *Adaptations; Feelings; Family stressors; Health; Personal responses; Experiences with services and professionals*. These provided insight into the complex responses

and effects of the young person's illness on family members. These themes are also captured in Figure 2.

[Insert Figure 2 here]

The illness transition for adolescents and young adults

Living with a life limiting condition formed a constant key transition for the adolescents and young adults, regardless of how long they had been living with the illness. The nature of the illnesses, ranged from general palliative care needs (Bousso et al., 2012, Wood et al., 2010), Duchenne Muscular Dystrophy (Abbott et al., 2012, Read et al., 2010), Cystic Fibrosis (Berge et al., 2007, Palmer and Boisen, 2008), Progressive Neurogenerative Disease (Bettle and Latimer, 2009), cancer (Flavelle, 2011, Kars et al., 2010, Kars et al., 2011b, Kars et al., 2011a), brain tumours (Freeman, et al , 2003,), spinal cord tumours (Freeman et al 2003) and Progressive Neurogenerative Illness (Rallison and Raffin-Bouchal, 2013). Moreover, some illnesses were unspecified, but were described as life limiting (Brennan et al., 2012, Cadell et al., 2012, Knapp et al., 2010, Menezes, 2010). This meant that there were mental, physical, emotional and spiritual adaptations that were necessary in order to live with their illness. Whilst some papers included the young person retrospectively thinking about the onset of diagnosis, illness transition was largely discussed in terms of the young person dealing with illness progression.

Illness trajectory

This sub-theme highlighted the ways in which the illness progression and experience affected the young person. This involved the deterioration caused by illness, and the adaptations that were needed to accommodate the changes brought about by illness. Constraints caused by illness and its progression was focused on in some studies, including how the illness impacted on aspirations for work (Abbott et al., 2012), participation in social activities (Berge et al., 2007), mobility and other physical, mental and emotional deterioration and impact on daily activities (Flavelle, 2011, Freeman et al 2003).

Inaddition, Palmer and Boisen's (Palmer and Boisen, 2008) study which focused on young adults with Cystic Fibrosis, revealed that their illness had brought about a sense that they were different from others, even in periods of good health.

Generally, perspectives from the adolescents and young adults were notably sparse within the included literature, with only six studies allowing space, in some form or another, for adolescents and young adults to disclose their experiences. This highlights a future potential research agenda, given that the young men featured in Abbott et al. (Abbott et al., 2012) spoke about appreciating the opportunity to talk about their illness and its trajectory. The other studies primarily focused on the perspectives of family members, which restricted the expansion of the *Illness trajectory* subtheme from the perspective of the adolescent or young adult.

Coping strategies

There was limited exploration of coping methods as reported by the adolescents and young adults themselves. The study looking at young men living with Duchenne Muscular Dystrophy (Abbott et al., 2012) found that they attempted to conceal their emotions in order to protect their parents. The adolescents and young adults often developed particular attitudes and took particular approaches to help them cope. This included focusing on the future to motivate compliance with treatment, as well as, adopting exercise to reduce treatment (Berge et al., 2007). In addition, accepting the illness as part of one's identity, lifestyle and/ or approaching illness through regular personal traits such as sarcasm (Berge et al., 2007, Flavelle, 2011, Palmer and Boisen, 2008), accepting support from relevant people and forums (Berge et al., 2007, Palmer and Boisen, 2008), participating in enjoyable activities (Bettle and Latimer, 2009, Flavelle, 2011, Palmer and Boisen, 2008) were also prevalent. Finally, putting things into perspective by looking at positive effects such as how illness had helped them develop a mature attitude (Palmer and Boisen, 2008) were identified. Flavelle (Flavelle, 2011) on the other hand, uses the theme of "Escape from illness" to describe ways in which the young person with cancer in her phenomenological study sought ways to distance himself from the pervading manner of his cancer. Strategies included video games, using humour, writing a journal, sleep and exploring different food.

Impact of illness transition on significant others

As seen in Figure 2, impact of illness transition largely related to family members such as parents and siblings, and the variety of effects was best described through six subthemes.

Adaptations

Many of the studies which focused on family members discussed some form of adaptations that had been made to accommodate the child with the life limiting condition and/or the overall situation. These sometimes involved lifestyle changes including house renovations (Bettle and Latimer, 2009), changes to employment arrangements (Cadell et al., 2012), sacrificing previous plans (Knapp et al., 2010), gaining expertise in their child's condition and care needs (Wood et al., 2010), and altering one's perception of time (Rallison and Raffin-Bouchal, 2013). This latter situation arose for family members of children with a Progressive Neurodegenerative Illness, who decided to live for each day as opposed to making plans for the future. This further resonated in families learning to focus on quality of life rather than the quantity of time the child had left to live (Rallison and Raffin-Bouchal, 2013). There were also emotional adaptations such as finding ways to keep a balance between needing to deal with having a poorly child and maintaining regular family dynamics and routines (Bousso et al., 2012), devoting efforts towards meaningful parenting (Kars et al., 2010, Kars et al., 2011b, Kars et al., 2011a), postponing grief while celebrating positive periods (Kars et al., 2011a), perceiving the child as strong (Bousso et al., 2012) and siblings compartmentalising home from school. Furthermore, in their study of the lived experiences of family members of children with illnesses, Wood et al. (Wood et al., 2010) described milestone events within phases of the disease trajectory, including moving from "normal" to "abnormal". However, there were challenges to adapting to emotional challenges such as coping with children dying before parents. This could be likened to existing within a duality of joy and sorrow (Rallison and Raffin-Bouchal, 2013). Part of adapting was

acceptance and this included siblings accepting caring as part of their identity (Brennan et al., 2012), adapting to a new “normality” (Wood et al., 2010), as well as, moments of “letting go” when parents accepted that their child’s cancer condition was terminal (Kars et al., 2010, Kars et al., 2011b, Kars et al., 2011a).

Feelings

Feelings which emerged in response to living and/or caring for a young person with a life limiting condition were centred around fear (Bettle and Latimer, 2009, Rallison and Raffin-Bouchal, 2013); uncertainty (Bettle and Latimer, 2009); sadness, grief and loss (Bettle and Latimer, 2009, Kars et al., 2011b), uncertainty and unpredictability (Bettle and Latimer, 2009); preserving hope (Betz et al., 2013, Bousso et al., 2012, Kars et al., 2010, Kars et al., 2011b, Kars et al., 2011a, Menezes, 2010), and the complexities of experiencing a duality of joy and sorrow (Rallison and Raffin-Bouchal, 2013).

Family stressors

Families faced financial burdens as a result of caregiving activities and adaptations to the home (Cadell et al., 2012, Monterosso et al., 2007, Read et al., 2010, Wood et al., 2010). Stressors were also impacting on family dynamics with members experiencing social isolation (Monterosso et al., 2007) and parents facing relationship strains (Berge et al., 2007, Bousso et al., 2012, Monterosso et al., 2007). Stress also materialised with regards to thinking about other children in the family who could not be focussed on as much and/or were kept sheltered (Monterosso et

al., 2007, Rallison and Raffin-Bouchal, 2013). Moreover, specific stressors were elaborated upon in some studies. Bousso et al.'s (2012) study (Bousso et al., 2012) on family members of children receiving home-based palliative care identified that following general struggles, stressors intensified once the terminal stage in the illness trajectory became more apparent. Bettle and Latimer's single case study (Bettle and Latimer, 2009) of the mother of a 16 year old young boy with a progressive neurodegenerative disease found that she experienced physical and emotional exhaustion due to having to attend to her son's illness needs, alongside, maintaining responsibility for other family members. However, the mother's coping strategies were described as drawing upon the strong, cohesive family system, and seeking information. Likewise, Brennan et al.'s study (Brennan et al., 2012) of siblings of children with life limiting conditions suggested that coping strategies saw the former use distraction, wishful thinking, resignation and social withdrawal in order to avoid stressors.

Health

A number of health implications were reported as being part of the impact of being related to a young person with a life limiting condition. These included mental health problems, such as depression, anxiety, stress and other emotional problems (Abbott et al., 2012, Bettle and Latimer, 2009, Bousso et al., 2012, Read et al., 2010). In Monterosso et al. 's study (Monterosso et al., 2007) which involved interviews with 134 parents of children receiving palliative and supportive care from community and hospital settings, half the parents reported that their mental health had suffered the

biggest effect. Physical health problems such as exhaustion (Monterosso et al., 2007) and fatigue (Knapp et al., 2010) were also described by family members.

Personal responses

Personal responses were commonly reported from mothers and siblings. For siblings, this included perceiving themselves as being different from the “norm” (Brennan et al., 2012), jealousy at receiving less parental attention (Berge et al., 2007), deterioration of school work (Freeman et al 2003),

Meola C,), and emotional difficulties (Read J et al., 2010). Positive impacts included having an enhanced desire to care and aspiring to have a career in the caring profession (Freeman et al 2013). The mother featured in Bettle and Latimer’s (2009) case study (Bettle and Latimer, 2009) described her “chronic sorrow” characterised by emotional responses to the child’s illness, including anger, sadness, fear and hopefulness. However, these responses are periodic and may lessen and/or intensify depending on the developing situation. One study (Bousso et al., 2012) found that mothers were more likely to see themselves as protective and were more prominent in caregiving activities such as attending health appointments. Conversely however, Flavelle’s analysis (Flavelle, 2011) revealed how, although the young adult’s mother wanted to be present throughout her son’s care, she struggled to deal with his affected leg. He noted that his father was more willing to get involved and described finding his father’s presence as comforting.

Experiences with services and professionals

The illness journey brought about interactions with services and healthcare professionals. Yet, some service provision and care approaches were reported to be problematic (Abbott et al., 2012). Positive services were acknowledged as implementing good coordination, future planning and allowing continuity through consistent relationships with professionals (Abbott et al., 2012). However, negative experiences were more commonly disclosed, which added to the stress experienced by family members. This negative outlook existed around lack of information or struggles around acquiring information (Cadell et al., 2012, Monterosso et al., 2007, Rallison and Raffin-Bouchal, 2013, Wood et al., 2010). Monterosso et al.'s study (Monterosso et al., 2007) found that parents of children who had non-cancer conditions faced more struggles with accessing appropriate information. Moreover, family member participants in Wood et al. 's study (Wood et al., 2010) revealed that there were some situations in which trust in professionals had to be re-negotiated following circumstances where it was felt the latter had made wrong decisions. In addition, the sibling participants in Freeman et al.'s 2003 study (Freeman, O'Dell, Meola 2003,) reported that they felt unprepared for death, due to sparse information being offered to them by professionals.

Developmental transition for adolescents and young adults

As recognised in the literature, the element that distinguishes this particular patient group from others is that adolescents and young adults are progressing into adulthood. This is encapsulated within the developmental transition. This review found that pertinent to this transition, were aspects around personal development. This led to a number of subthemes which could all be related to how the young

person sought to develop themselves and move into, or further into, the phase of adulthood.

Personal development- Employment/ Education/ Training

This aspect of the developmental transition was widely reported in the studies. Whilst pursuit for work was aspired to, there was recognition of how illness exacerbated the difficulty in securing employment (Abbott et al., 2012). Aspirations for educational development and employment were affected by illnesses, with many adolescents and young adults anticipating barriers based on their condition (Abbott et al., 2012). Moreover, Palmer and Boisen's study (Palmer and Boisen, 2008) showed that future educational aspirations were dictated to some extent by illness considerations. These adolescents' and young adults' Cystic Fibrosis diagnosis meant that therapy equipment and support needed to be convenient. Thus location of college needed to be within a convenient distance of the support centre. Moreover, the nature of the illness also influenced the type of accommodation that could be aimed for with some young adults choosing to remain living with parents as opposed to student accommodation. Student accommodation needed to be planned so that therapy equipment could be accommodated, which was not feasible when the accommodation was a room share. However, all the adolescents and young adults in this study reported that they were currently in employment with professional posts, and many worked on a full time basis (Palmer and Boisen, 2008).

As some of the adolescents and young adults included in these studies were adolescents, there was also exploration of issues regarding school and school work. Bettle and Latimer's single case study (2009) describe the determination of the mother is resulting in her son being enrolled into a specialised school program, which incorporated a personal school assistant. In Freeman et al.'s (2003)

study on children with brain or spinal cord tumours keeping up with school work was reported as a heightened problem for affected children during hospitalisation.

Personal development- Relationships and Socialisation

The pursuit of socialisation was recognised as a key milestone for young people in the adolescent and young adult age range, but was accepted that it was difficult to achieve due to the challenges of the illness (Bettle and Latimer, 2009, Freeman et al 2003).

Moreover, Abbott et al. (Abbott D et al.,) highlight that older participants with Duchenne Muscular Dystrophy felt that they had less opportunity to establish social networks, and desired to socialise beyond family members. Young adult participants with Cystic Fibrosis in Palmer and Boisen's (2008) study disclosed that they considered their social life to be active and on a par with their peers who did not have a life limiting condition. However, they clarified that this was only possible through careful organisation in order to also accommodate healthcare activities. In addition, these participants acknowledged some limitations in their approaches to socialisation such as avoiding smoke-filled settings. Some adolescents and young

adults were reticent about disclosing their illness in new relationships (Palmer and Boisen, 2008).

In addition, Flavelle's study (2011) in relation to a young adult's diary disclosures reveal a different type of relationship by highlighting how his relationship with God progressed to include trust in Divine protection and regular Bible reading. Moreover, this young person describes how present relationships altered with his preference for his father to spend time with him over his mother as illness progressed. This young person had a younger sibling and revelled in his role as a big brother. However, following illness progression, he discusses the need to protect his sibling from the illness prognosis.

Personal development- Future planning

The notion of future planning encompassed life goals and aspirations such as relationships and finding cures for their illnesses. Young female adults with Cystic Fibrosis (Berge et al., 2007) used the goal of starting a family as a motivator for complying with treatment. Moreover, in the study of the young person with cancer (Flavelle, 2011), the participant shared his desire to have future intimate relationships, but on the other hand, revealed that he did not want to burden anyone else with his illness. Other than relationship goals, some studies revealed participants' hope in finding a cure for their illness and/ or beating their illness (Berge et al., 2007, Flavelle, 2011). Beyond specific goals, the female participants in Berge et al.'s study discussed how they had accepted their Cystic Fibrosis as being part of

their identity and this aided them in their general decisions and plans for the future. Likewise, Flavelle (2011) highlights how the young person with cancer had also accepted his illness and began to think about how his personal effects should be distributed after his death. Acceptance of illness was also influential in future planning for the participants in Palmer and Boisen's study (2008) as all formed hopes and goals, while analysing their circumstances in a positive way, reiterating that things could be a lot worse.

Personal development- Independence

Independence was a key element and priority for participants and was described as a typical adolescent goal (Flavelle, 2011, Palmer and Boisen, 2008). Illness-related independence was achieved through treatment decisions and taking responsibility for health (Flavelle, 2011, Palmer and Boisen, 2008). Independence as relating to healthcare responsibility was already strong for participants in Palmer and Boisen's study (Palmer and Boisen, 2008), but once living away from parents, more initiative was employed. Moreover, pursuit for independence began to transpire in other areas beyond health, for example finance, diet and more general decision making. Challenges to achieving independence included feelings of not being listened to (Palmer and Boisen, 2008) and the awareness that peers without illness were probably able to achieve better independence(Palmer and Boisen, 2008). However, Palmer and Boisen's (2008) participants also perceived themselves to be at a higher maturity level compared to their peers, as they had been pushed into adulthood because of their illness.

Discussion

Concurrent transitions

This review has revealed the pertinent issues around the developmental transition for adolescents and young adults with life limiting conditions, which revolve around personal development. Also, of significance is the illness transition, which encompasses deterioration, limitations and modified aspirations as part of the trajectory. These concurrent transitions make such adolescents' and young adults' needs complex. Previous work has tended to discuss aspects of personal development within "transition to adulthood", a broad transition which also depicts the movement to adult services (Abbott et al 2012). To avoid confusion, we have been explicit to name these aspects of personal development that are present when moving to adulthood as the developmental transition, which we feel more faithfully encapsulates these experiences. Past research has also failed to reveal the illness transition as it occurs concurrently with experiences inherent in the developmental transition. Furthermore, this review has drawn attention to the impact of the illness transition on family members, highlighting the effect on their physical, mental and emotional health.

The concurrent transitions and notable impact of illness on family members suggests that the complex environment of the young person needs to be taken into account when addressing multiple needs. Bronfenbrenner's ecological systems theory (Bronfenbrenner, 1979) supports this notion by postulating that all aspects and levels of a child's environment are influential in how they grow and develop. This includes the immediate relationships and organisations, which are present in the child's life,

such as family members, healthcare professionals and prospective employers (microsystem); the interactions between the different members of the child's microsystem (mesosystem); the wider agencies and organisations that do not have direct contact/ interactions with child, but still impact on his/her life (exosystem); and finally the widest system that although remote, has a great influence over the child, for example, the economy, cultural values and attitudes (the macrosystem). This perspective highlights the need to take into account not just the young person, but also their interactions and associations with people in their lives. Therefore, Bronfenbrenner's work enables insight into the needs of these young adults as they navigate through transitions and development. Our review findings suggest that the dynamics of the mesosystem need strengthening to ensure that the young person can be adequately supported during their transitions. Moreover, according to Bronfenbrenner's understanding into the chronosystem (Bronfenbrenner, 1986, Hosek et al., 2008), environmental changes need to be taken into account over time. Normative transitions appear to include the developmental transitions such as puberty, education and marriage, whilst non-normative transitions encompass the unexpected and often negative events, such as illness, divorce and death. With regards to the concurrent transitions highlighted in this review, normative transitions relate to the personal development experiences and goals inherent in the developmental transition. Non-normative transitions relate to the illness transition events. Both of these impact on adolescents' and young adults' environment and development, and therefore need equal attention in the context of the young adults having a life limiting condition.

Bury's theory of biographical disruption (Bury, 1982) is also relevant here, and highlights how illness causes everyday structures present in life to be disrupted. This leads to the person re-thinking their life course as issues such as pain and death become immediate possibilities. Increasing dependency on others and the presence of uncertainty add to the disruption. Human agency may be pivotal in enabling the person to navigate their life course in the face of illness (Sanders et al., 2015). Our review suggests in the presence of illness and the illness transition, it is then the developmental transition, which motivates a desire for continuity, facilitates the formation of aspirations, and stimulates human agency. For some adolescents and young adults, aspirations at this point may involve a modified version compared to their peers, to take into account their illness, but nonetheless, they are still held and pursued.

Implications for research and practice

Future research should endeavour to further investigate the health, educational and social needs that appear to be pertinent. Moreover, insights from adolescents and young adults themselves are severely lacking, which means an underrepresentation of their valid experiences and concerns. With further regard to the range of life limiting conditions being considered, some young adults with these conditions may not be able to articulate their views clearly due to their associated disabilities. They may require the assistance of parents, siblings, other carers, or technology to support their inclusion. Nevertheless, it is very important that their views are heard, as their views may differ from those of their more able (but still life-limited) contemporaries. The focus on proxy views needs to be taken with caution, since they

may not be representative of the actual adolescent or young adult. Further exploration of the impact of illness on the whole family unit is also necessary.

Limitations

Search strategies, however comprehensive, may fail to reveal relevant studies. Due to resource limitations, we were unable to include studies not published in English. Any form of critical appraisal risks are based on subjective criteria, which might mean that some relevant papers were excluded.

Conclusion

This review is original in that it has synthesised the findings from different studies to provide the evidence around pertinent transitions for adolescents and young adults with life limiting illnesses. The two concurrent transitions of illness transition and developmental transition highlighted the ways in which adolescents and young adults with life limiting conditions have particularly complex needs. In the face of illness, the concurrent developmental transition stimulates a desire for continuity and pursuit of age-appropriate goals and aspirations. These may be modified to accommodate the life limiting condition. In addition, the impact of illness transitions on family members establishes some of the ways in which family members may also require support during the illness trajectory and beyond. More research around the perspectives of young adults would further expand understanding into the two concurrent transitions.

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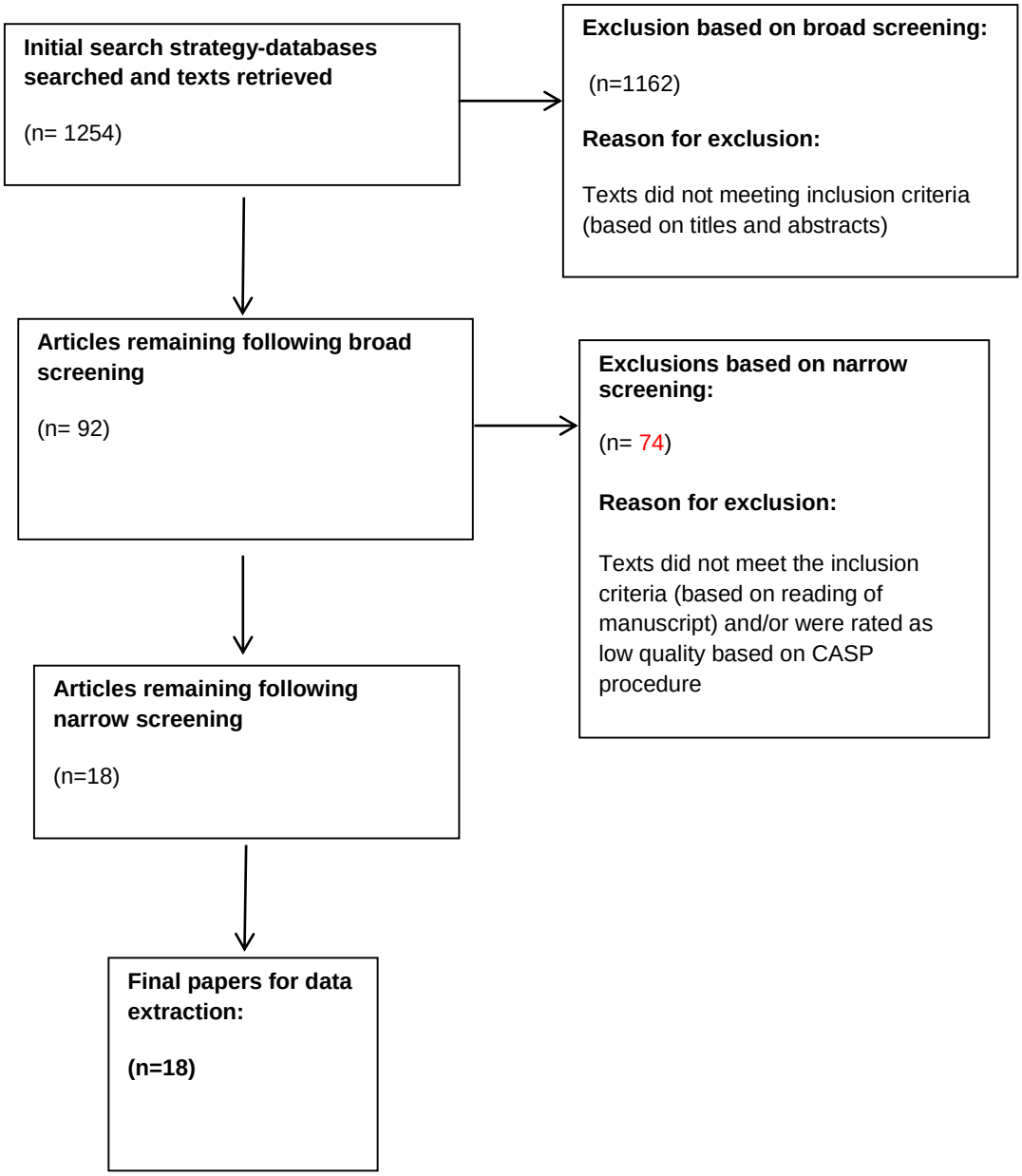
Figure 1: PRISMA flow diagram

Figure 2: Key transitions for adolescents and young adults and impact of significant others



Legend: The PRISMA flow diagram shows the stages of narrowing down papers, including reasons for excluding papers.

Table 1: Inclusion and exclusion criteria for review

Inclusion criteria	**Exclusion criteria**
1. Adolescents and young adults (young people) aged between 10-25 years, with life limiting condition (as categorised by ACT), family members or care staff, or other professionals (including volunteer staff) involved with these young adults 2. Relevant to life transitions and situations of the young person and/or their family 3. Empirical research (qualitative or quantitative) published since the year 2000, to ensure recent relevance to current clinical services	1. Young adults with chronic or developmental disorders which are not intrinsically life-limiting (e.g. diabetes endocrine, or learning disorders). 2. Service or care transitions, as sole focus 3. Retrospective studies where the life issues and views of the young adult are not represented 4. Care/service evaluations 5. Technological or legal issues as sole focus 6. No English language translation available.

Table 2: MEDLINE search strategy

No.	Term
1.	young adult/
2.	adolescent/
3.	adolescent health services/
4.	family/
5.	siblings/
6.	caregivers/
7.	health personnel/
8.	volunteers/
9.	young adult$. tw
10.	adolescen$.tw
11.	adolescent health service$.tw
12.	famil$.tw
13.	family member$.tw
14.	sibling$.tw
15.	carer$.tw
16.	healthcare personnel.tw
17.	health care worker$.tw
18.	healthcare staff.tw
19.	teaching staff.tw
20.	teacher$.tw
21.	volunteer$.tw
22.	young person.tw
23.	young people.te
24.	or/1-23
25.	palliative care/
26.	terminal care/
27.	life expectancy/

28.	palliative care.tw
29.	terminal care.tw
30.	life expectancy.tw
31.	end of life.tw
32.	life limit$.tw
33.	life shorten$.tw
34.	or/25-33
35.	life change events/
36.	adaptation, psychological/
37.	social adjustment/
38.	family relations/
39.	parent-child relations/
40.	family health/
41.	life change event$.tw
42.	adaptation, psychological.tw
43.	social adjustment$.tw
44.	family relation$.tw
45.	parent-child relation$.tw
46.	family health.tw
47.	life experience$.tw
48.	life issue$.tw
49.	family system$.tw
50.	life transition$.tw
51.	family dynamic$.tw
52.	or/35-51
53.	24 and 34 and 52
Limiters: date of publication 20000101-20140103; English language; human not animal	

Table 3: Overview of included studies

Reference	Participants/ diagnosis or condition	CASP SCORE	Methods	Summary of findings/ transitions
1. Abbott D., Carpenter J., Bushby K. (2012)Transition to adulthood for young men with Duchenne Muscular Dystrophy: research from the UK, Neuromuscular Disorders, 22: 445-446	Young men living with Duchenne Muscular Dystrophy (DMD), parents and siblings Young men were >15 yrs; mean age 19.6 yrs	Moderate	Postal survey of parents (*N*=38) who have a son with DMD Individual interviews with 40 families consisting of the young men with DMD, their parents and siblings (*N*=102)	**Illness transition** - *Coping strategies* > The young men found it hard to stay positive tried to protect their parents by not showing their emotions. - *Dealing with service and professionals* ***Illness transition – Impact on parents*** - 80% of parents reported depression and anxiety, according to the Hospital Anxiety and Depression Scale (HADS). - Negative effects of problematic services, including

				confusion and anxiety - Parents felt demotivated when healthcare professionals focused on deterioration in the young men. **Developmental transition** - *Personal development* *- education/ employment* *-Personal development* - *relationships* networks .
2. Berge JM, Patterson JM, Goetz D, and Miller C. (2007) Gender differences in young adults' perceptions of living with cystic fibrosis during the transition to adulthood: a qualitative investigation, Families, Systems and Health, 25(2): 190-203	17 young adults with Cystic Fibrosis (Females *N*=11; Males *N*=6).	high	4 focus groups (Males 16-18 yrs; Males 19-21 yrs; Females 16-18 yrs; Females 19-21 yrs. The data underwent grounded hermeneutic analysis	**Illness transition** - *Illness trajectory* away from parents; - differences between males and females - *Coping strategies* > issues regarding treatment and support as well as identify ***Illness transition- impact on families***

				Developmental transition *- Personal development - relationships* > *-Personal development - future planning* >
3. Bettle A., and Latimer MA. (2009) Maternal coping and adaptation: a case study examination of chronic sorrow in caring for an adolescent with a progressive neurodegenerative disease, Canadian Journal of Neuroscience Nursing, 31(4): 15-21	Mother of one young adult (16 yrs) with progressive neurodegenerative disease of unknown origin	moderate	Single case study of mother's experience. A family systems model was used when exploring maternal coping and adaptation.	**Illness transition** *- Illness trajectory* > Developmental transitions and constant health care needs > Developmental and cognitive deterioration and losses have been slow and gradual > *- Coping strategies* ***Illness transition- impact on mother*** - **Developmental transition**

				- Personal development - *Relationships* > *-Personal development* *- Education* .
4. Bousso R., Misko MD., Mendes-Castillo AMC., Rossato LM. (2012) Family management style framework and its use with families who have a child undergoing palliative care at home, Journal of Family Nursing, 18(1): 91-122	11 families- 14 members in total (parents, aunt, grandmother). Children aged 3-16 years receiving palliative care at home	moderate	1 interview with parent(s) and extended family members (either with a single person, or as a family group) using Family Management Style Framework	***Illness transition- impact of parents*** -
5. Brennan C., Hugh-Jones S., and Aldridge J. (2012) Paediatric life-limiting conditions: Coping and adjustment in siblings, Journal of Health Psychology, 18 (6): 813-824	31 siblings aged between 5 and 6 yrs of children with a life-limiting condition	high	Longitudinal mixed method study	***Illness transition- Impact on siblings*** - - a
6. Cadell S., Kennedy K., and Hemsworth D. (2012) Informing Social Work Practice	47 interviewees, plus 273 returned questionnaires (from parents, grandparents,	high	Postal questionnaires and content analysis of semi-structured interviews	**Illness transition- Impact on family members** -

	Through Research With Parent Caregivers of a Child With a Life-Limiting Illness, Journal of Social Work in End-of-Life & Palliative Care, 8(4): 356-381	step or foster parents of adolescents/ young adults aged 19 yrs or younger).		(either individual or with another family member).	
7.	Flavelle SC. (2011) Experience of an Adolescent Living With and Dying of Cancer, Archives of Pediatric Adolescent Medicine, 165 (1): 28-32	Young adult (Ed) with cancer- aged 16 yrs	moderate	Phenomenological study of personal diary entries	**Illness transition** - *Illness trajectory* with his sister *- Coping mechanisms* **Developmental transition** - *- Personal development - achievements* > Pride drawn from achievements *- Personal development - relationships* > *- Personal development - future planning*
8.	Freeman K., O'Dell C., Meola C.	Children with brain or spinal cord tumours	moderate	Survey	**Illness transition**

	(2003) Childhood Brain Tumors: Children's and Siblings' Concerns Regarding the Diagnosis and Phase of Illness, Journal of Pediatric Oncology Nursing, 20(3): 133-140	and their siblings from 40 families. Mean age of affected children was 15 yrs; Mean age of siblings was 17 yrs			*- Illness transition- Experiences of services and professionals* > *- Illness transition- Illness trajectory* > ***Illness transition- Impact on siblings*** > **- Developmental transition** *- Personal development - Education* > Inadequate support in keeping up with school work was reported as an issue during hospitalisation *- Personal development - Relationships* > Difficulties in socialisation came to the

				fore during remission
9. Kars MC., Grypdonck MHF., Beishuizen A., Meijer-van den Bergh EMM. And Delden JJM. (2010) Factors Influencing Parental Readiness to Let Their Child With Cancer Die, Pediatric Blood Cancer, 54, 1000-1008	44 parents of 23 children with incurable cancer at the end of life phase	high	Multi-centre qualitative study using single or repeated interviews	***Illness transition- Impact on parents:*** - Moving between "Preservation" and "Letting go"
10. Kars MC., Grypdonck MHF. And van Delden JJM. (2011a) Being a Parent of a Child With Cancer Throughout the End-of-Life Course, Oncology Nursing Forum, 38(4): E260	42 parents of 2 children with incurable cancer, cared for at home	high	Single and repeated interviews subjected to inductive thematic analysis	***Illness transition- impact on parents*** - - Managing the change for the worse - Being with the dying child - Protecting and upholding the parent-child relationship. - Strategies for coping with the end of life
11. Kars MC., Grypdonck MHF., de Korte-Verhoef MC., *et al.* (2011b) Parental	44 parents of 23 children with incurable cancer	high	Single and repeated open interviews with parents	***Illness transition- impact on parents*** - Feelings of loss,).

experience at the end-of-life in children with cancer:'preservation' and 'letting go' in relation to loss , Supportive Care in Cancer, 19, 27-35				- Forced to acknowledge loss when parents learn that treatment has failed-r and sometimes even co-exist
12. Knapp CA., Madden VL., Curtis CM., Sloyer P. and Shenkman EA. (2010) Family Support in Pediatric Palliative Care: How Are Families Impacted by their Children's Illnesses? Journal of Palliative Medicine, 13 (4): 421- 426	85 parents who had children with life limiting illnesses, and were enrolled in a publically funded pediatric palliative care programme	high	Cross-sectional telephone survey, using the main outcome measure of the Impact on Family (IOF) scale (a 15 item measure, which looks at social and personal strain using a 4 point Likert scale).	***Illness transition-impact on family*** - .
13. Menezes A. (2010) Moments of realization: life-limiting illness in childhood-perspectives of children, young people and families, International Journal of Palliative Nursing, 10(1): 41- 47	11 children and young people affected by life-limiting conditions and their parents and siblings (39 participants from 10 families)	high	Qualitative research- case study using interviews. Parents were allowed to use of visual materials to help convey their family life.	**Illness transition** *-Illness transition-Illness trajectory* > and the consequences ***Illness transition-impact on family*** - ct that the child had time to live

14. Monterosso L., Kristjanson LJ., Aoun S., and Phillips MB. (2007) Supportive and palliative care needs of families of children with life-threatening illnesses in Western Australia: evidence to guide the development of a palliative care service	134 parents of children receiving palliative and supportive care in community and hospital settings, and 20 service providers	high	Interviews	**Illness transition:** *Illness transition- Illness trajectory* - ***Illness transition- Impact on family*** - Emotional, financial and physical burdens – exhaustion. - - Struggles faced from lack of clear information about their child's condition, conditions, practical aspects of care were less likely to be met, - Parents also faced concern over skills of carers in end of life care - Siblings faced emotional impact from their brother or sister's condition but parents often shielded the former from caregiving

				tasks to help them ***Illness transition- impact on service providers*** -
15. Palmer ML., and Boisen LS. (2008) Cystic Fibrosis and the Transition to Adulthood, Social Work in Health CareSocial Work in Health Care, 36(1): 45-58	7 young adults with Cystic Fibrosis (2 male, 5 female). Age range-20-26 yrs	moderate	Qualitative content analysis of individual in-depth interviews	**Illness transition** *Illness transition - Illness transition- coping mechanisms* > Organisation and planning allowed time for social activities as well as healthcare schedules > Perceiving therapy as a positive component of their regime > CF itself allowed participants to put things into perspective and dissuaded them from worrying about other things > > Support from loved ones was appreciated,

				> Employing a positive outlook and recognising the qualities that they had developed because of their CF **Developmental transition** - *Personal development class incomes and lifestyles"* (p48). > - *Personal development - Independence* > > Diet needed to be based on healthier selections, which ended up being more costly > - *Personal development – Education* > - *Personal development - Relationships and socialisation*

				- Personal development *- Future planning*
16. Rallison LB. and Raffin-Bouchal S. (2013) Living in the In-Between: Families Caring for a Child With a Progressive Neurodegenerative Illness, Qualitative Health Research, 23(2): 194-206	27 members from 6 families (parents, siblings, grandparents, and people considered to be family); One child with PND	high	Phenomenological study using individual or group in-depth interviews; observation of children with PND	***Illness trajectory-Impact on families*** > Holding predictability and unpredictability > Hope and despair > Feeling of parental failure > Fear and subsequent exhaustion > Adjusting to new concept of "time"-
17. Read J., Kinali M., Muntoni F. and Garralda ME. (2010) Psychosocial adjustment in siblings of young people with Duchenne muscular dystrophy, European Journal of Paediatric Neurology, 340-348	46 siblings; 39 parents/ main carers; 25 teachers	high	Various questionnaires; semi-structured interview	***Illness transition-Impact on siblings and others*** - -
18. Wood F., Simpson S., Barnes E., Hain R. (2010) Disease trajectories and	36 family members (parents, siblings, foster carers) from 26 families	high	Thematic content analysis of individual semi-structured	***Illness transition-Impact on family members*** >

	ACT/ RCPCH categories in paediatric palliative care, Palliative Medicine, 24(8): 796-806	Ages of ill children not made explicit		interviews, describing lived experiences of families	

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20. Berge JM, Patterson JM, Goetz D, and Miller C. (2007) Gender differences in young adults' perceptions of living with cystic fibrosis during the transition to adulthood: a qualitative investigation, Families, Systems and Health, 25(2): 190-203	17 young adults with Cystic Fibrosis (Females *N*=11; Males *N*=6).	high	4 focus groups (Males 16-18 yrs; Males 19-21 yrs; Females 16-18 yrs; Females 19-21 yrs. The data underwent grounded hermeneutic analysis	**Illness transition** - *Illness trajectory* away from parents; - differences between males and females - *Coping strategies* > issues regarding treatment and support as well as identify ***Illness transition- impact on families***

				Developmental transition *- Personal development - relationships* > *-Personal development - future planning* >
21. Bettle A., and Latimer MA. (2009) Maternal coping and adaptation: a case study examination of chronic sorrow in caring for an adolescent with a progressive neurodegenerative disease, Canadian Journal of Neuroscience Nursing, 31(4): 15-21	Mother of one young adult (16 yrs) with progressive neurodegenerative disease of unknown origin	moderate	Single case study of mother's experience. A family systems model was used when exploring maternal coping and adaptation.	**Illness transition** *- Illness trajectory* > Developmental transitions and constant health care needs > Developmental and cognitive deterioration and losses have been slow and gradual > *- Coping strategies* ***Illness transition- impact on mother*** - **Developmental transition**

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23. Brennan C., Hugh-Jones S., and Aldridge J. (2012) Paediatric life-limiting conditions: Coping and adjustment in siblings, Journal of Health Psychology, 18 (6): 813-824	31 siblings aged between 5 and 6 yrs of children with a life-limiting condition	high	Longitudinal mixed method study	***Illness transition- Impact on siblings*** - - a
24. Cadell S., Kennedy K., and Hemsworth D. (2012) Informing Social Work Practice	47 interviewees, plus 273 returned questionnaires (from parents, grandparents,	high	Postal questionnaires and content analysis of semi-structured interviews	**Illness transition- Impact on family members** -

Through Research With Parent Caregivers of a Child With a Life-Limiting Illness, Journal of Social Work in End-of-Life & Palliative Care, 8(4): 356-381	step or foster parents of adolescents/ young adults aged 19 yrs or younger).		(either individual or with another family member).	
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34. Rallison LB. and Raffin-Bouchal S. (2013) Living in the In-Between: Families Caring for a Child With a Progressive Neurodegenerative Illness, Qualitative Health Research, 23(2): 194-206	27 members from 6 families (parents, siblings, grandparents, and people considered to be family); One child with PND	high	Phenomenological study using individual or group in-depth interviews; observation of children with PND	***Illness trajectory-Impact on families*** > Holding predictability and unpredictability > Hope and despair > Feeling of parental failure > Fear and subsequent exhaustion > Adjusting to new concept of "time"-
35. Read J., Kinali M., Muntoni F. and Garralda ME. (2010) Psychosocial adjustment in siblings of young people with Duchenne muscular dystrophy, European Journal of Paediatric Neurology, 340-348	46 siblings; 39 parents/ main carers; 25 teachers	high	Various questionnaires; semi-structured interview	***Illness transition-Impact on siblings and others*** - -
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	ACT/ RCPCH categories in paediatric palliative care, Palliative Medicine, 24(8): 796-806	Ages of ill children not made explicit		interviews, describing lived experiences of families	