Transition in Cystic Fibrosis: An International Experience

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8.1 Introduction

Cystic fibrosis (CF) is a life-limiting, autosomal recessive, genetic disease primarily affecting the sodium channels within the body making it difficult for salt and water to travel between cells. Consequently, mucus within the body is thick and viscid leading to disease and injury in the affected organs. Although the effects of CF on the respiratory and gastrointestinal tracts are the main cause of morbidity and mortality, CF can also cause endocrine, reproductive, metabolic and psychological disease. While survival is constantly improving with new medication and early treatment, over time there is a relentless decline in health leading to premature death. Treatment for CF is daily, time consuming and complex; it is understandable therefore why adherence to treatment remains an issue, particularly during the adolescent years (Quittner et al. 2014). Taking responsibility for managing the demands of CF is a significant step towards independence and adult care.

Cystic fibrosis was previously known as a disease of childhood with those affected unlikely to live into early adulthood. However, through early and aggressive treatment of respiratory infection, an increase in the use of mucoactive therapy, changes in pancreatic enzyme replacement and multidisciplinary care for children and adults, survival has improved (Burgel et al. 2015). Registry data from 2017 from both the UK Cystic Fibrosis Trust (2018) and the American Cystic Fibrosis Foundation (2018a) suggests that a child born in 2017 has a median predicted survival of 47 years of age, this does not take into account new therapies.

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United Kingdom CF registry data (UK Cystic Fibrosis Registry 2018) further shows fewer deaths of children and young adults (≤19 years), with only ten deaths reported in 2017 (out of 4816 ≤ 19 years). As the birth rate remains constant, many countries around the world are starting to report more adults than children with CF, consequently the need for thoughtful, robust transition programmes from paediatric to adult care are essential. Burgel et al. (2015) report that the adult CF population in Europe is likely to increase by 75% by 2025, driven primarily by a healthier paediatric population transitioning into adult services. This predicted population increase presents a challenge to existing CF services throughout Europe which will need to adapt in order to provide care to a larger population.

Cystic fibrosis is described as an orphan disease—affecting fewer than 200,000 people globally, current data suggests that there are approximately 44,000 people living with the disease in Europe and 30,000 in the USA (Cystic Fibrosis Foundation 2018a; Orenti et al. 2018). The benefit of this relatively small global patient population means that the CF clinical community readily shares research, service development and care guidelines, routinely inviting people with CF and family members to be involved in decision-making endeavours. These collective efforts often mean that aspects of care are relatively standardised across the world.

This chapter will address issues pertaining to transition directly related to CF; additionally transition policies from various countries will be discussed. Data from an international survey of CF transition programmes will be presented with findings demonstrating the variation among policies and practices. An overview of the current transition trends and practices within CF will be presented.

8.2 Common Key Features of International Transition Programmes in Cystic Fibrosis

There is broad, international agreement of the transition process from specialist paediatric CF centre care to specialist adult CF services. However, because of varied national guidelines, geography, lack of specialist adult care and lack of resources, the details differ. The first section of this chapter highlights common key areas of discussion: age of transition, parental involvement, educational programmes, formal transition processes, as well as communication and co-ordination. Key features of CF programmes in the UK, Australia, Canada and the USA are presented.

Age at transition: Although it is universally accepted that transition means moving from specialist services provided in childhood to complimentary ones in adulthood, there is little agreement in the literature of an ideal age (Gravelle et al. 2012). Many national guidelines for CF care have been led by consensus recommendations which have been used to develop local practice. There is therefore variance between practice in different countries and even centres within the same country (Gravelle et al. 2012). Age guidelines of Australia, USA, Canada and the UK are presented below.

In Australia, transition is discussed with young people and their families at diagnosis with more in-depth discussion beginning at the age of 12 years, followed by referral to the adult centre at 18 years of age (Reid et al. 2008). However, the guidelines offer some flexibility, stating that young people may transition earlier on a case-by-case basis if they are deemed mature enough to move to adult services at an earlier stage (Reid et al. 2008). Additionally, it is recognised that transition of young people who are at the stage of palliative care or transplantation creates challenges for both the adult and paediatric teams, as well as the young person and their family. These situations require significant multidisciplinary team communication and planning to facilitate a successful transition (Towns and Bell 2011).

Various sources of transition information from America suggest that transition for adolescents with CF takes place between the ages of 17 and 21 years, although it has been suggested that transition may really take place between the ages of 14 and 30 years (Gravelle et al. 2012). In the American CF population, it has been argued that by encouraging young people who are reluctant to engage in the transition process to move their care, the CF multidisciplinary team (CF MDT) either risks refusal to engage or disengagement from healthcare systems altogether (Flume 2009). There is, however, a counter argument suggesting that by not encouraging transition to adult services there is a risk of disengagement from healthcare altogether, as young people may feel they have outgrown the paediatric service and do not know how to engage with an adult service (Flume 2009).

Guidelines for transition in Canada recommend transition to adult services at the age of 18 years. Most paediatric centres in Canada (91%) refer to an adult CF centre at the age of 18 years; however, a minority (9%) struggle to find an adult centre to refer to, often because of remote or rural locations (Gravelle et al. 2012).

European recommendations are more general as they offer advice to a wide and varied population within Western and Eastern Europe. Resources are hugely varied and in many countries adult CF care services do not exist. Cystic Fibrosis Worldwide (2018) suggests that in countries where CF is less common in the general population many children may be treated for symptoms but will die without being diagnosed. In countries such as El Salvador, India and Bulgaria the average age of death is 15 years old; therefore, transition services are yet to be developed. The European Standards of Care do not comment specifically on the age at which young people should transition and, as our survey highlights, there is variance in the age at which young people move to adult CF services across Europe (Conway et al. 2014). In the UK, guidelines suggest that the transition process should start at a young age but that formal discussions should start at least a year before moving to adult care. Timing of this move should be flexible in accordance with need, but broad age limits are recommended to be between 14 and 18 years (Transition Fact Sheets, Cystic Fibrosis Trust 2013).

Involving parents: Transition may appear to be focused on the young person, primarily the individual who is moving from one healthcare system to another. However, it is also a process in which the young person's parents/carers and wider

support network are also involved. In CF, this is particularly pertinent as the adolescent and parents will have to spend many years in close association with the paediatric team. This is acknowledged internationally with all countries recognising the long-term paediatric relationship and the necessity for providing adult care to these young people, balanced with the needs of the parents/carers. Although global healthcare transition guidelines provide limited guidance pertaining to parental involvement, recommendations presented below are based upon available evidence.

Parental involvement and support should not be underestimated in CF and often aids in the overall success of transition (Towns and Bell 2011). Healthcare transition occurs at a time of wider transition in a young person's life as it relates to education, social and emotional changes, the development and exploration of new relationships and beginning of autonomy and independence in managing healthcare needs (Conway et al. 2014).

Generally, the literature suggests that parents/carers often have higher levels of anxiety regarding transition than young adults. This is supported by Van Staa et al. (2011), who found that young adults with CF going through transition often have a "wait and see" approach, whereas parents feared leaving a safe environment and felt a sense of loss at the prospect of their child moving to adult services. Parents of children with CF are used to frequently contacting the paediatric healthcare practitioners and often know the team very well (Van Staa et al. 2011). Consequently, having previously been central to their child's management with control over treatment and communication with their child's care provider, many parents struggle to build a trusting relationship with healthcare practitioners in adult services. Flume et al. (2001), further suggest that parents may not want to relinquish control and may doubt their child's abilities to manage their own healthcare. Of note, the legal age of majority differs between countries and within countries, and although most recognise 18 years, this can be as low as 14 years and up to 21 years (United Nations Human Rights: Office of The High Commissioner 1990).

Whether parents' reluctance to trust their children with healthcare-related decision making is true or not, this concern often leads them to describe a lack of involvement and a feeling of dismissal by both their adult child and the adult team. It is important to note, however, that young people often value their parents' support and assistance with medications, prompting them to complete treatment, advocating for them and assisting while they are experiencing a respiratory exacerbation. Coyne et al. (2018) suggest that the transfer of responsibility for management of the young adults' condition is a gradual process and one that should be allowed to take some time.

Available American guidance on transition does not give any indication of the level or manner in which parents should be involved in the transition process (Yankaskas et al. 2004). Australian standards suggest that the transition "should

engage the young person with CF and their family in a positive way". European standards of care make recommendations regarding parental involvement and support, which can be offered while their child with CF is very young; however, it does not make any recommendation regarding the involvement of parents throughout the transition process (Conway et al. 2014). The UK Cystic Fibrosis Trust (2013) produces an information fact sheet for parents that explains why transition is necessary, the differences between paediatric and adult care, the changes they as parents and their child can expect, and what their roles as parents may be throughout the process.

Educational programmes: Several educational programmes that have been developed in Australia, the USA and Canada will be discussed. Guidance from Australia recommends that as transition approaches, a greater emphasis on self-care skills and developing independence should be highlighted by the paediatric CF MDT. Towns and Bell (2011) advocate for early introduction of self-management materials in adolescence to aid in the development of knowledge and skills; encouraging the young person to gradually become more independent. There should be easy access to information about adult services including information about the adult MDT staff and any specialist advice which may be offered, for example, employment/higher education, fertility and assisted conception, transplantation and other issues encountered in adulthood with CF. Cystic fibrosis MDTs should tailor education to the concerns raised by young people and their families around transition, and that members of either the adult or paediatric team should be easy to contact regarding any queries.

The American Cystic Fibrosis Foundation does not give any specific information on transition regarding education programmes (Cystic Fibrosis Foundation 2018b). However, Okumura et al. (2014) describe a programme to improve transition from paediatric to adult services within their CF population in San Francisco, California. As described by Okumura et al., education is key and a number of educational materials have been developed by the CF MDT to increase knowledge of CF and assist in the development of independent management and advocacy abilities. These resources, in the form of checklists, guides and notebooks are introduced to all patients at the age of eight years old, with the expectation that families will bring these to each appointment and use them to act as prompts for discussions relevant to self-management and transition during consultations. Through discussion and working within the developmental framework, the paediatric team can determine whether a young person is developing independence and self-management skills. These resources aim to help the children and young adults develop coping skills, increase autonomy and promote strategies to increase adherence.

American transition guidance also supports a programme called CF R.I.S.E which is designed to aid young adults transitioning to adult services. This programme supports the development of skills needed to be responsible for

self-care, developing independence and the provision education (CF RISE 2019). The programme also provides timelines of goals to be achieved, with a set of tools which can be used with parents/carers and healthcare providers to evaluate appropriate levels of knowledge and skills to manage CF independently (CF RISE 2019).

Authors from the paediatric CF centre in British Columbia (Canada) have described a multidimensional transition programme (On Trac) which includes a clinical pathway, collaboration with the adult CF MDT and a tool to measure transfer readiness. Similar programmes are used by other provinces, for example, 'On my way' and 'You're in charge' (Gravelle et al. 2012). Additionally, acknowledging the vast distances some families must travel to access CF care and the CF MDT, the Canadian CF charity 'Cystic Fibrosis Canada' provides a virtual education programme for young people with CF and their parents which includes content on transition.

Neither the European standards of care nor the UK Cystic Fibrosis Trust mention specific transition education programmes, although local programmes have been developed in some paediatric CF centres and only used in-house, therefore, not published. The Cystic Fibrosis Trust, however, does recommend that specialist adult CF centres provide information about the transition service including clinical arrangements, in-patient facilities, the names of the CF MDT and directions to the centre (Cystic Fibrosis Trust 2013).

Formal transition programmes: Young people report feeling concerned about meeting a new team when there is a lack of information about the clinicians, concerns about the quality of care and apprehension about the infection control protocols used (Boyle et al. 2001; Coyne et al. 2017). As findings reveal, anxiety among young adults can be significantly reduced by meeting the adult team and visiting the adult CF centre in anticipation of the transfer of care. A systematic review by Coyne et al. (2017) found that young people moving from paediatric CF services into adult care worried about leaving their existing care team, which they may have known their entire lives. In this section, the status of formalised transition programmes that have been developed worldwide are presented.

Transition advice from both the American and Australian patient advocacy groups does not include information about formalised CF transition programmes (Cystic Fibrosis Foundation and Cystic Fibrosis Australia). However, transition processes for professionals and patients are commented on, although the method or manner of transition is not directly discussed (Reid et al. 2008). Flume (2009) proposes that although many studies advocate for the inclusion of joint paediatric and adult clinics at the time of transition, this is often not a practical solution for centres within the USA which may be a great distance away from the referring centre, even crossing state boundaries. Instead, it is suggested that the young person have an informal tour of adult CF centre with a member of the adult team (Flume 2009).

Towns and Bell (2011) found that despite the lack of national transitional guidelines in Australia, some states such as New South Wales have created state-wide transition programmes which are used by all adolescents with chronic health requirements within certain geographical boundaries. The introduction of liaison between paediatric and adult CF MDTs was suggested as part of the transition process as it helped in communication and collaboration. Towns and Bell noted that, through working closely together, paediatric MDTs had a greater awareness of the differences in adult healthcare and could discuss concerns raised by young adults and their families in a supportive manner.

In Canada, Gravelle et al. (2012) report in 2007 only a small proportion (22%) of Canadian CF centres had a formal transition process, the majority of which had been developed by the CF nursing teams, based upon existing literature and protocols. Approximately a quarter of centres established a transition programme utilising a joint clinic or "graduation clinic" involving members of both the adult and paediatric MDTs. Subsequently, when the data were reviewed in 2011, most Canadian CF centres had developed their own transition programmes and practices.

The UK Cystic Fibrosis Trust does not mention formal educational programmes; however, the National Institute for Health and Care Excellence (The National Institute for Clinical Excellence [NICE] 2016) has formulated general recommendations regarding the transition from paediatric to adult health services across all specialties. Recommendations include the following: a) early discussion and planning beginning at age of 13 years; b) transition should take place at a time of relative stability in the young person's life and c) young people should be actively involved in planning and making decisions about where they would like their adult care to be provided. These guidelines also introduced the concept of a key worker who oversees the transition process and provides information and support to the young person and their family. The guidelines suggest that support should be in place both before and after transition for a minimum of six months and longer if required.

Ready, Steady, Go, a transition programme based upon the NICE guidance (www.uhs.nhs.uk/readysteadygo) has been adopted widely in paediatric CF centres in the UK. This programme provides a structure to ensure all young people and their carers are appropriately supported through the transition process and can be used for all young people with a long-term condition aged 11 years and beyond. Materials include transition policies, implementation guides, questionnaires and information about the process.

Communication and co-ordination: Strategies for fostering communication have been recommended by programmes internationally. The guidance from Australia, the Netherlands and the USA are reviewed below. Australian guidance on transition recognises the importance of communication between paediatric and adult MDTs in co-ordinating transition (Reid et al. 2008). It is also suggested

that successful transition depends upon the cooperation of both paediatric and adult MDTs, who should meet regularly to discuss who will be moving to adult services within the following year. Efforts should also be made to include local doctors and hospitals who may be involved in the young person's care. A summary of care from birth to transition should be provided for the adult MDT including information regarding CF and non-CF related complications, psychological well-being, intravenous access and hopes for the future. The guidelines recommend a clear plan that includes contact information for paediatric and adult providers during the transfer period between the final paediatric appointment and first adult clinic visit, in the event that the young person needs assistance (Towns and Bell 2011).

The role of a transition co-ordinator is briefly discussed in the Australian guidance, stating that the co-ordinator should meet young adults and their families in the year prior to moving to an adult CF centre, and the young adult should have the opportunity to visit the adult CF centre (Reid et al. 2008). Towns and Bell (2011) support this, saying that transition co-ordinators were found to help the transition process by offering support to both young people and their families, as well as the entire clinical team. Additionally, they were able to provide education and training, smoothing the transition process.

Dutch data published by Van Staa et al. (2011) suggests that in some cases there is little co-ordination between adult and paediatric services in the Netherlands. Transition services which integrate both adult and paediatric MDTs allow for greater collaboration, increased communication and an easier transition. American researches by Okumura et al. (2014) advise that fostering a culture of dual responsibility between the adult and paediatric CF MDT is essential when developing a transition programme. Australian guidance proposes that transition policies and guidelines are agreed upon and developed by both adult and paediatric CF MDTs who work together regularly (Towns and Bell 2011). This is supported by American literature which suggests regular discussion between paediatric and adult teams leads to the development of a shared approach to CF management (Flume 2009).

Canadian guidance from Gravelle et al. (2012) suggests that although the entire MDT should be involved with transition, transition programmes benefit from having one member of staff taking charge and co-ordinating this process. Gravelle et al. (2012) suggest that in a survey conducted in the USA less than fifty percent of CF centres identified a member of staff responsible for co-ordinating transition despite this being indicated in their national guidance (Yankaskas et al. 2004). Further, in the majority of CF centres, the role of transition co-ordinator was the CF specialist nurse. The UK NICE guidelines (2016) recognise that transition between services is often a complicated process which can involve multiple agencies. If a co-ordinated approach is not employed this may result in poor communication, decreased engagement and continuity of care may be affected. Teams involved in the transition process should be clear about their roles and responsibilities in the process.

8.3 International Transition Survey

Based on this review of the literature, a short questionnaire about transition from paediatric to adult CF care was circulated during the Nurses Special Interest Group meeting at the European Cystic Fibrosis Society Conference in 2018. Twenty-five replies were received from twelve countries representing Scandinavia, Western Europe, Eastern Europe, North America, Australasia and South Africa. Respondents were from both urban and more rural centres, some requiring patients and families to travel long distances for many hours to access care. Each country follows national guidelines; however, geography and population determine local practice.

The questionnaire was designed to gather data on key features of transition programmes established in the CF services of the 12 countries. Questionnaire items included the age of the first discussion about transition and the consequent age of transition, parental involvement in the process, use of educational programmes, communication between the paediatric and adult CF multidisciplinary teams, existing formal transition programmes, co-ordination of the process and resources used throughout the transition and transfer of care. These items were selected from the CF-related transition literature by a group of paediatric and adult CF clinical nurse specialists.

Although individually, national guidelines for the transition of children to adult CF care exist in various countries around the world, there does not appear to be any global consensus about the ideal age of the first discussion or actual transition of care. Most respondents reported 14–18 years (n = 19, 76%) as the most frequently identified ages to start the conversation (Fig. 8.1). Recognising transition is a lifelong process; several respondents described the importance of early discussion and education. This approach was aimed to allow children and their families to become

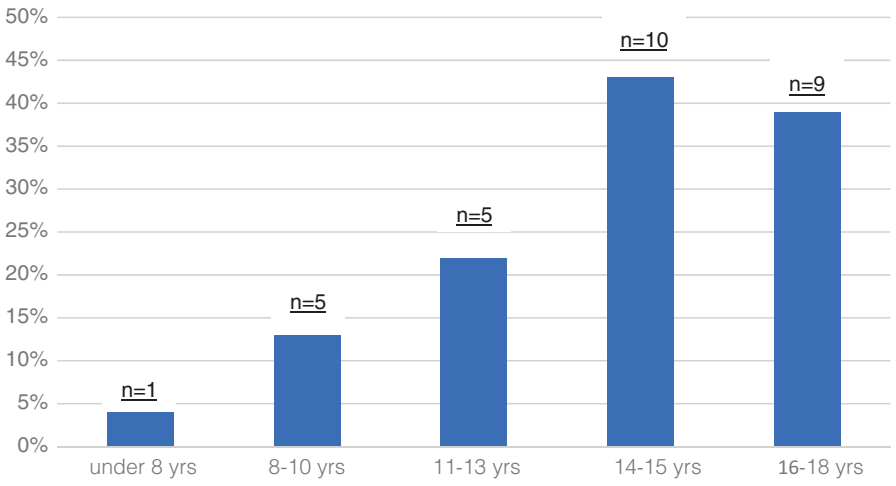

Fig. 8.1 At what age do you start discussing transition to adult services? (Respondents reported more than one age group)

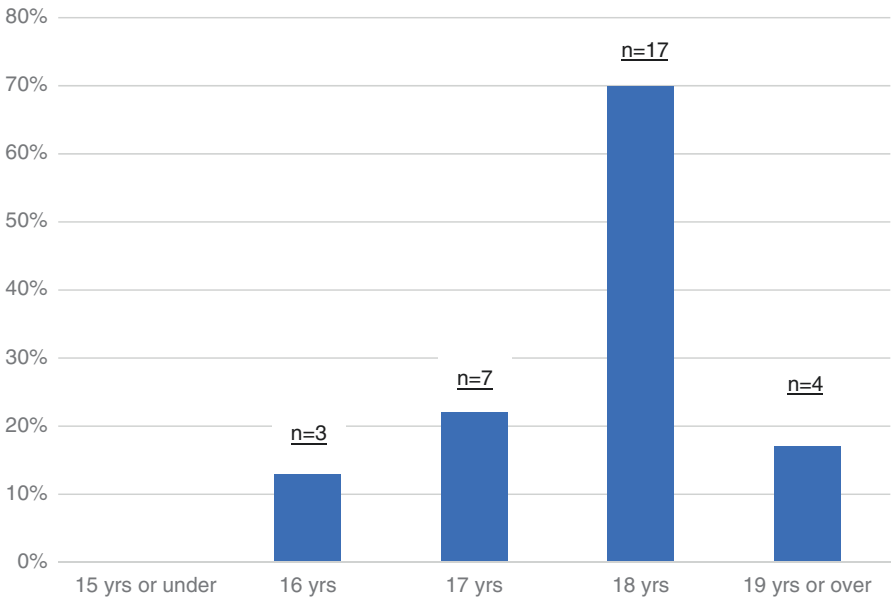

Fig. 8.2 At what age do patients transition into the adult service? (Respondents reported more than one age group)

empowered in active decision making about care. This policy was reflected in 24% (n = 6) reporting that they started transition discussion under the age of 10 years. Most respondents (n = 17, 68%) reported 18 years as the ideal age for transition to adult services (Fig. 8.2). All respondents (n = 25, 100%) indicated that parents are involved in the transition process.

As discussed earlier in this chapter, education and support from a member of the CF MDT are key recommendations when planning transition. Respondents were therefore asked about the use of educational programmes and key workers to support the process. Education directed at transition was used in just under half of the centres (n = 11, 44%), with just over half of the programmes using a specific key worker (n = 15, 60%) to support children and their families through the process.

Historically, transition from paediatric to adult CF care has been organised by the paediatric team. However, developing a formal transition programme has increasingly become a joint paediatric and adult CF multidisciplinary initiative. Respondents were asked if they used a formal transition programme with the majority agreeing (n = 16, 64%) and only four saying they did not follow any programme. The rest (n = 5, 20%) were currently developing a programme. Respondents indicated the initiation time frame of their transition programme: 12 programmes (48%) five or more years; 7 (28%) for 1–4 years; and 3 (12%) less than a year.

As has been discussed, communication and co-ordination are key to a successful transition programme and process for the patient and their family (Flume 2009). Respondents were asked about methods of communication between the paediatric

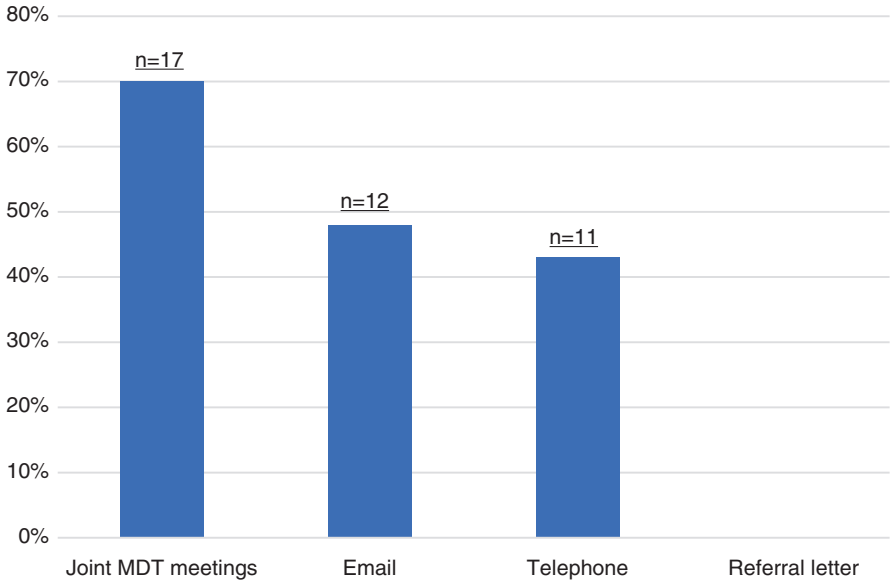

Fig. 8.3 How do you communicate between adult and paediatric teams to discuss transition? (respondents used more than one method)

and adult CF MDTs, responsibility for co-ordination and any formal monitoring of patients and families who disengage during the process. Most respondents (n = 17, 68%) reported that they hold joint paediatric and adult multidisciplinary meetings, with email and telephone as adjuncts to communication. No centre relied on referral letters (Fig. 8.3). Fifty-six percent (n = 14) of respondents identified the clinical nurse specialist as the co-ordinator, although there was occasionally more than one co-ordinator. One respondent identified co-ordinators who sat outside the multidisciplinary team—family doctors and the clinic co-ordinator (Fig. 8.4). Half (n = 13, 52%) of respondents monitor patients and families who disengage from the transition process with the majority (n = 13, 48%) of this co-ordination carried out by clinical nurse specialists.

Finally, respondents were asked about the different resources used by CF MDTs to support patients and families through the process. Although the majority (n = 16, 64%) use printed resources—booklets or transition ready questionnaires, many augment this with information on websites (n = 8, 32%) and a selection of phone or tablet applications, DVDs or videos (n = 7, 28%). A number of respondents described developing technical innovations including a virtual reality tool to allow patients and families to experience an outpatient visit to the adult CF service.

Surveys can only ever represent the opinions of the sample respondents, however, in this instance, surveys were returned from different continents and therefore may go some way in presenting a more representative overview of transition in CF worldwide. Areas where there were no replies are unfortunately not represented in this survey; they include America, the Middle East and Central and South-East Asia.

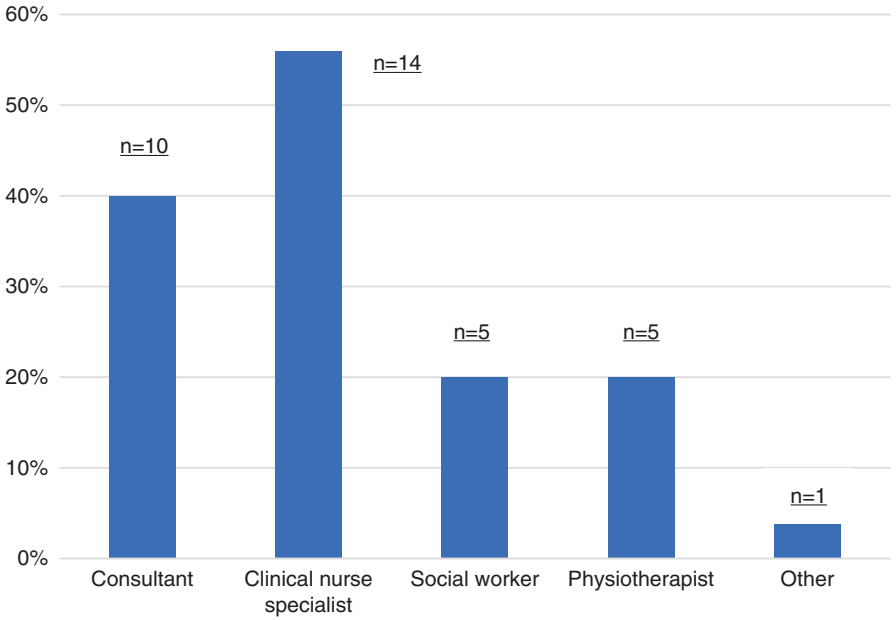

Fig. 8.4 Who co-ordinates your transition service? (respondents used more than one professional)

The incidence of CF is low in the Middle East and Central and South-East Asia with no formal speciality, paediatric and adult CF centres established to date (Cystic Fibrosis Worldwide 2018).

The literature describes various national guidelines, advice and consensus agreement which has been discussed earlier in this chapter. The results of this survey appear to support the themes presented throughout the discussion of the guidelines reviewed.

8.4 Recommendations

The key discussion points raised in this review chapter are summarised below. Although there is no international, formal consensus about these points, it appears that most countries appear to follow a broadly accepted transition journey that incorporate published guidelines.

When to Start Talking about Transition: Literature around transition in CF, guidelines and standards of care suggest that early conversations with both children and their parents/carers about transition from paediatric to adult care are important. The survey findings showed that most of the CF centres follow a policy of introducing the topic between the ages of 14 and 18 years. In the UK and Australasia, discussions tend to begin earlier at 12–13 years, with transition taking place between the ages of 16 and 18 (Bourke and Houghton 2018). In the USA, CF MDTs appear to begin discussions later, with a consequent later transition to the adult service, between

the ages of 17 and 21 years. Sawicki et al. (2018) reviewed CF transition practices between 2007 and 2013 and found that although almost 70% of young adults transitioned to adult centres between the ages of 18 and 21 years, there was still some inconsistency of age at transfer, which varied between 14 and 56 years old. In contrast, comments from the survey suggest that some CF centres ($n = 4$) believe that to normalise transition as part of growing up, conversations should start from diagnosis and be re-visited at least annually. Based upon the evidence presented in this chapter, the recommendation is proposed for initiation discussions about transition.

In acknowledging these variations, it seems that conversations about transition should be normalised at a very young age. However, more formal discussion and preparation should take place at least from the age of 12 years.

Age of Transition: Based on the information reviewed to date, transition usually takes place between the ages of 16 and 21 years and beyond (Bourke and Houghton 2018; Sawicki et al. 2018). There are several issues influencing transition including available resources, accessibility of adult CF centres, local and national guidelines concerning the appropriate age a young person can be transferred to an adult ward, and the reported needs of the young person and their parent/carers.

Taking the above points into consideration, the informal consensus opinion for the ideal age of transition is 18 years.

Parental Involvement: The information we have reviewed in this chapter certainly suggests that parents/carers should be involved in the transition process, and that globally, CF centres recognise the importance of including parents throughout the journey. This is evidenced from the survey wherein 100% of teams involve parents. However, although the degree to which parents are involved varies greatly around the world, the disparity in international practice highlights that although there is general agreement, there is no consensus on the most beneficial way to engage with parents (Iles and Lowton 2010).

General opinion recognises the importance of involving parent/carers throughout the process, even if the level of inclusion varies.

Educational Programmes: Our survey suggests that just under half of CF services (44%) provide formal education about transition and preparation for adulthood with CF. These educational materials vary between checklists and guides designed to prompt the healthcare professional to provide education on a certain topic; use of methods for determining the young person's level of understanding and giving practical advice regarding the adult service (Connett and Negra 2018; Gravelle et al. 2015).

Based on this information, we would suggest at the very least, that practical information regarding the adult CF service should be provided. This should include information regarding location of the service, clinic schedules and frequency of reviews, provision for urgent review, location and layout of the ward and facilities available, and the contact details for the team. Additionally, formal education programmes designed to develop autonomy and knowledge should be available.

Formal Transition Processes: This review shows an increase in the numbers of CF centres that have developed and implemented formal transition programmes over the last few years. As evidenced in the literature and based upon our survey results, geography, particularly distances between paediatric and adult services, as well as the lack of adult CF services in some countries can make setting up formal transition programmes more difficult. The service models vary as some transition programmes typically involve young people and their parents/carers through graduation clinics and transition ready clinics, whereas other centres use a transition key worker acting as a link between the paediatric and adult CF services (Towns and Bell 2011. Gravelle et al. 2015). Alternatively, some centres use specific transition education programmes such as 'On Trac' in Canada, 'Ready Steady Go' in the UK, and a transition readiness scale in the USA (Gravelle et al. 2015. Connett and Negra 2018. Lapp and Chase 2018).

We suggest that a formal programme, agreed upon by both the paediatric and adult teams, allows for a more supportive process for the young person and the parent/carer. This may involve following a readiness programme or incorporating a key worker to co-ordinate the process.

Communication and Co-ordination: Communication and co-ordination is clearly an essential component of the transition process, although less than 50% of survey respondents have a specific team member co-ordinating transition. Where co-ordinators are in place, the role often falls to the CF nurse specialist with assistance from the wider MDT. There is no agreement on the frequency of paediatric/adult team meetings, although this may differ depending on the size of the centre and the number of young people being transitioned. Individual teams decide on the communication process which works for them, from face-to-face meetings, to using available technology for communicating over greater distances.

Paediatric and adult CF centres need recognised pathways of communication to ensure a successful transition for each young person. This process is often led by a co-ordinator, which commonly is a CF specialist nurse.

8.5 Summary

As healthcare professionals, we need to decide what makes transition successful in our services. As treatment for CF has improved over the last few decades there has been a marked rise in survival, with most children born with CF living well into adulthood. This creates challenges as existing CF services must adapt to this increasing demand for adult services. In the future, we can expect growing numbers of young adults accessing adult CF programmes; this will require CF teams to have robust processes in place to facilitate their transition. Most CF services are currently in the process of adapting or developing their transition programmes to facilitate this rise in the CF population.

It is beneficial for healthcare professionals to begin conversations about transition early, hopefully the subject could be introduced to parents at diagnosis and with the subject of transition discussed more formally with the young person beginning at 12 or 13 years of age. Paediatric and adult teams should communicate regularly about patients who are beginning the transition process, using shared processes and protocols for a transition programme for which both teams share a vision. Transition needs to involve both the paediatric and adult multidisciplinary teams, with the opportunity for a formal handover to the receiving team prior to the patient completing the transition journey.

Increased collaboration and communication between teams leads to a smoother transition process for young people and their families. Transition programmes have been found to benefit from one person co-ordinating the transition process often, but not exclusively, this is the role of a nurse specialist. The transition co-ordinator is responsible for the following transition services: a) assessing transition readiness; b) providing support and education to young people and their families throughout the transition process; c) giving information regarding the similarities and differences of the adult service; and d) monitoring for patient disengagement after transition.

We also need to ask ourselves what is important for young people and their families. Transition can be daunting for young people and their families who are likely to have developed positive and trusting relationships with their paediatric healthcare providers over many years. Research suggests that young people may find it easier to adapt to this change as they are going through a process of transition in many areas of their lives at this time and may have a more relaxed approach to transition than their parents (Van Staa et al. 2011). Flume et al. (2001) found that parents are often more apprehensive regarding transition as they have been central to their child's care and management over the preceding years. It is important to recognise that for patients and their families, transition should be a gradual process, throughout which they should be supported by the transition co-ordinator and multidisciplinary team.

Young people and their families should be provided with information regarding what to expect throughout the transition process. Materials should be available explaining how the adult service works and, although the format of this information varies between different transition programmes, information is

commonly provided in a written format or online. Young people should be supported and encouraged by the MDT to increase their knowledge regarding their treatment and develop independence not only with treatment management but all aspects of daily living. For those who are reluctant to engage in the transition process, the reasons for this hesitancy should be explored by the transition co-ordinator allowing for any concerns to be addressed. Their concerns may need the assistance of a counsellor to address them. Young people and their families should have the contact details for the transition co-ordinator, allowing easy access to information if questions arise between appointments; research suggests that informal visits of the adult CF centres lead to decreased anxiety among young people and their families (Coyne et al. 2017).

Transition from paediatric to adult care in a life-long, life-limiting disease is challenging for the young person, their family and the clinical teams, both paediatric and adult. Close, trusting relationships develop between the child, parents and CF MDT which can be hard to give up. However, despite an ambition to provide a great service, there are two important challenges to consider. Distance between centres and resources are challenges as young people may travel long distances to attend routine CF appointments because of geography and/or lack access to resources. The lack of adult centres, particularly in some Eastern European and Asian countries is an issue that the international CF clinical community is trying to address, although this will take time. Distance and geography mean that the timing of appointments may need to be customised; however, a number of centres where this is an issue use telemedicine, including the use of video monitoring or virtual consultations, particularly when patients are known to be clinically stable. Pre-transition preparation, meeting the adult team and working through education programmes may be possible through this technology.

Lung transplantation or complex disease is a unique challenge facing some young people with CF. We can expect that most young people who transition to an adult CF service will be reasonably well, with a relatively well-preserved lung function and good nutrition. However, for a minority this may not be the case as some may have severe disease with associated co-morbidities or be navigating the lung transplantation referral process. Transition of a young person with rapidly declining health or who may need to consider a transplant referral process presents several challenges. Difficult decisions around the appropriate place of referral may lead to a potential two-step transition: first the transfer to paediatrics transplant services followed by referral to adult CF services. Alternatively, options could include staying in paediatric services with a well-known MDT for end of life care, or transition to a specialist adult CF service where the CF MDT are unknown but have the skill and knowledge to manage the young person. Consideration of the views of the young person and their family must be taken into account; however, the skills of managing end of life care in the paediatric team, the availability of transplant services and distance should also be discussed. Taylor et al. (2006) suggest that young people thinking about lung transplant at the time of transition may feel a sense of rejection from the paediatric team as transitioning to transplant can have negative connotations. There is a risk that

families may feel overwhelmed by the volume of information they are receiving, while meeting new team members from multiple care teams during this period of transition.

8.6 Conclusion

When considering transition, it is undoubtedly important that a formal process should be developed and implemented by local teams. Communication between teams is essential and will lead to a streamlined and smooth transition process. Paediatric and adult CF MDTs should agree and individualise an age that they aim to transition each patient by, although this varies internationally but is usually around the age of 18 years. Education and support should be provided to all those going through the transition process, the rationale for transition should be explained, for example, the adult team's expertise in supporting education, employment, fertility and treatment management.

Transition continues to be an important aspect of care in CF; most young people cared for by CF MDTs today can expect to progress into adult services as relatively well young adults. The demand, therefore, for a smooth, co-ordinated, supportive multidisciplinary transition process will continue. This chapter highlights the challenges facing the CF clinical community in supporting young people and their families through the transition process and gives an overview of current international practices.

8.7 Useful Resources

United Kingdom

NICE shared learning database: Organisation: University Hospital Southampton NHS Foundation Trust. January 2017

- Implementing transition care locally and nationally using the 'Ready Steady Go' programmewww.nice.org.uk/sharedlearning/implementing-transition-care-locally-and-nationally-using-the-ready-steady-go-programme
- www.uhs.nhs.uk/readysteadygo

Cystic Fibrosis Trust Fact Sheets

- Transition from paediatric to adult care: a guide for young people.
- Transition from paediatric to adult care: a guide for parents.
- Transition from paediatric to adult care: a guide for commissioners, hospital and clinical teams.
- www.cysticfibrosis.org.uk/the-work-we-do/publications/factsheets-and-information-packs

Royal Brompton Hospital transition animations

- https://www.youtube.com/watch?v=tt6jtLfxoak&feature=youtu.be
- https://www.youtube.com/watch?v=m3k89EqaT6w&feature=youtu.be

United States

- CF RISE – Responsibility. Independence. Self-Care. Education.
- https://www.cfrise.com/

Canada

- On Trac: http://ontracbc.ca/

Australia

- Sir Charles Gairdener Hospital, Western Australia - Moving on to Charlies.
- CF Transition Group Working Party Sir Charles Gairdener Hospital (2009).
- http://files.aussiehome.com/cmsFiles/651/TRANSITION%20BOOKLET.pdf
- Trapeze: A supported leap into adult health: The Sydney Childrens Hospitals Network http://www.trapeze.org.au/

Ireland

- Transition information and videos for young people with a long-term illness (www.Steppingup.ie). This web site provides information, resources, and videos of transition stories; to assist in thinking about transition, then planning and making a transition to a new health service.

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