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Published in:
Breathe

Publication status and date:
Published: 01/09/2022

DOI (link to publisher):
[10.1183/20734735.0157-2021](https://doi.org/10.1183/20734735.0157-2021)

Document Version
Publisher's PDF, also known as Version of record

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Citation for the published version (APA):
Office, D., & Heeres, I. (2022). Transition from paediatric to adult care in cystic fibrosis. *Breathe*, 18(3), Article 210157. <https://doi.org/10.1183/20734735.0157-2021>

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Transition from paediatric to adult care in cystic fibrosis

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Shareable abstract (@ERSpublications)

Transition to adult services should be discussed with young people with CF and their families from at least early adolescence. There should be an allocated transition key worker to support the young person and their family throughout the process. <https://bit.ly/3aKfMgB>

Cite this article as: Office D, Heeres I. Transition from paediatric to adult care in cystic fibrosis. *Breathe* 2022; 18: 210157 [DOI: 10.1183/20734735.0157-2021].

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Received: 25 Jan 2022
Accepted: 31 May 2022

Abstract

In the decades since cystic fibrosis (CF) was first clinically defined in the 1930s, there have been many advancements in the treatment and management of this disease. Initially it was considered a disease of childhood where the majority of those affected died before reaching adolescence. Now, through advancements in management and treatment, the vast majority of those affected will live into adulthood. Therefore, paediatric and adult CF services must collaborate to ensure that young people and their families experience a positive and supportive transition into adult services. Key aspects of transition will be discussed, including when to begin the transition process, who should coordinate this and how the transition process should be structured. Challenges of the transition process and potential pitfalls when transition does not run smoothly will also be discussed, as well as tools that may be used to support a positive transition for young people and their families.

Educational aims

- To familiarise readers with factors that make the transition process positive.
- To make suggestions regarding the application of the transition process.
- To highlight factors which may impact on the success of the transition process and the risks associated with disengagement at the point of transition.
- To discuss tools which can be used by care teams to ensure a smooth transition process.

Introduction

Cystic fibrosis (CF) is a chronic life-limiting disease caused by a sodium channel disorder influencing the transport of salt and water within the body. The primary cause of morbidity and mortality is chronic bacterial respiratory infection and inflammation as viscid secretions build up in the airways [1, 2]. However, CF is a multisystem disorder with gastrointestinal, endocrine, hepatic and reproductive comorbidities causing additional complications. In previous decades CF was known as a disease of childhood, with the majority of those affected not surviving into adulthood [3]. Thankfully, over time effective treatments have been developed and the majority of those born today can expect to live well into adulthood [3]. Additionally, the recent development and initiation of gene-modulator medications, available for up to 90% of the CF population, has enabled healthcare teams to provide highly effective therapies targeted at specific gene defects [4]. Therefore, over time many young people with CF will transition to adult services in relatively good health.

The literature suggests a 75% increase in those transitioning between paediatric and adult CF services in Europe between 2015 and 2025 [5]. This increase is mainly due to improvements in treatment in combination with lower paediatric mortality. Many countries around the world are now reporting greater numbers of patients in adult CF services compared with paediatric services [2, 5]. UK registry data report



that over 60% of those with CF receive treatment in adult care [6]. It is, therefore, imperative that CF care providers have robust transition services in place. This review discusses key points to consider in relation to the transition process in the young CF population, including factors which will have a positive influence on the transition process, suggestions for implementing transition in practice, factors which can affect the success of the transition process for young people and tools which may be useful.

Key points to optimise the transition process

When should transition begin?

It is generally accepted that transition discussions should take place from at least early adolescence to normalise the process of growing up and developing independence and self-management skills. Adolescence can be defined as the phase of life stretching between childhood and adulthood, and encompasses elements of biological growth and major social role transitions. Rather than age 10–19 years, a definition of 10–24 years corresponds more closely to adolescent growth and popular understandings of this life phase and would facilitate extended investments across a broader range of settings [7]. Adolescence is also a unique stage in human development and an important time for laying the foundations of good health. Adolescents need opportunities to meaningfully participate in the design and delivery of interventions to improve and maintain their health [8]. Some CF centres discuss transition with parents on an annual basis, over time the child becomes increasingly involved and the conversations become more directed towards them. The age at which transition occurs differs between countries and occasionally within the same country. Populations and societies function differently around the world and as such, the appropriate age for transition can vary. In a recent international survey of transition in CF the ages ranged from 16 years to 21 years [9]. Regardless of the chosen age, local teams should agree an age at which they aim to transition young people within their patient cohort.

Multidisciplinary care

Paediatric and adult CF teams who work together regularly should agree a shared set of goals and guidelines related to transition for young people moving between their services. Shared working across teams and becoming dually responsible allows for an effective transition. Defining the exact age of transition can be difficult as countries and health systems accept young people into adult care at different ages. However, pre-transition preparation does not need age definition as it takes place before the move to an adult centre regardless of age. Paediatric and adult multidisciplinary teams (MDTs) should meet regularly to discuss young people who will be transitioning between their services [10]. However, in some cases this may not be possible, for example if the young person is moving away to university or if the family relocate to another part of the country. Teams in the Netherlands have successfully used virtual meetings to transition patients between centres where patients have moved great distances, this is an effective mode of communication for CF teams who may not transition patients between their centres on a regular basis. The paediatric team should provide a comprehensive written handover to the adult team, often referred to as the young person's transition document. This document should summarise details of diagnosis particularly the method of diagnosis (*e.g.* newborn screening), sweat chloride values and genotype (which is necessary when considering access to modulators). Additionally, a summary of care provided from birth to the point of transition should be included, containing information regarding physical and mental health, complications both related and unrelated to CF, social circumstances, and information pertaining to the individual's future aspirations in relation to education and work [11].

Meeting the adult team

It is widely reported in the literature that young people and their families may experience anxiety and concern at the prospect of transitioning from paediatric services which have been the primary care providers for their entire lives. However, there is evidence that parental anxiety may supersede that of the young person who may have a more *laissez-faire* attitude to transition [12]. Commonly cited concerns include meeting new team members, anxiety regarding changes in quality of care between services, a much larger adult CF service with many more patients and worries regarding infection control [13]. Meeting the adult CF team prior to transition can significantly decrease anxiety and concern, as it offers the opportunity for both the young person and their family to ask questions and discuss concerns with the adult care providers [13]. In turn, through open discussion young people and their families can be reassured about the similarities and differences between adult and paediatric care and how support will be provided throughout the transition process. Meeting with young people and their families enables the adult team to talk about expectations during the transition process, including developing independence and practicing CF-related self-management skills [9]. Engaging with the adult team prior to transition is also an opportunity for the adult team to provide information and educational resources, both written and electronic, to help facilitate the transition process.

Young people should be encouraged to take an active part in the transition process, advocating for themselves and making decisions such as where they would like their adult care to be delivered. It is important that young people are able to make an informed decision about where to have their future care, many paediatric CF services run joint transition clinics with a number of adult providers, these are commonly available on one or more occasion prior to transfer to adult care. At these clinics young people can meet members of the adult team alongside familiar members of the paediatric team during joint consultations [14]. Joint transition clinics allow young people and their families to meet different adult teams before making a final decision regarding where their adult care will be based. Some young people will choose to attend the adult centre in the same hospital as their paediatric care, others may choose to move to an adult centre which is more geographically appropriate. The reported benefits of transition clinics are improved preparedness of young people and their families, and enhanced joint working between members of the paediatric and adult MDTs.

Coordination of the transition process

In the UK, the National Institute for Health and Care Excellence (NICE) published guidance regarding transition for use in all healthcare settings in which young people move from paediatric to adult services [15]. Recommendations include discussion of transition with young people (and their families) at an early age to normalise the process and encourage them to consider this as a positive step towards adulthood and independence. Where possible, transition should take place at a time of stability in the young person's life in terms of disease progression, social circumstances and education [15].

A further recommendation suggests that young people and their families could benefit from one point of contact, or a specific team member known as the transition key worker (TKW). A TKW assists young people and their families through the transition process providing individualised support for a minimum of 6 months pre- and post-transition [13, 15]. The TKW meets the young person and their family annually from the age of 14 years to answer questions regarding transition, provide reassurance and support, and aims to develop a trusting relationship until the young person adjusts to the culture of the adult centre and settles into the service [16]. Following transition, the TKW continues to provide continuity as a main point of contact, providing details so that contact can be made independently if there are questions or concerns between hospital visits. Further, the TKW supports adjustment to care in the adult service, attends/reviews progress at the first few adult clinic appointments, supports throughout the first admission in adult services and attends MDT meetings regarding the young person, such as those with school or social services, and ensures the young person attends appointments and remains engaged [8, 17].

Developing independence

When helping young people prepare to move to adult services it is important for them to become confident when discussing their symptoms with the members of the CF MDT. The paediatric MDT should encourage the young person to practice discussing their health prior to transition, perhaps with a paediatrician or a member of the MDT they feel comfortable with. Parents can be involved in this process, for example they may wish to remain in the room, observing but not actively taking part in the discussion. An advantage of this is that the parent remains with the young person throughout the consultation and can prompt the young person to discuss any health concerns they may have forgotten to mention or rejoin the conversation at the end.

Over time, as the young person develops confidence they are encouraged to talk to the team without their parents throughout their consultation. The parents can then rejoin the consultation, with the young person summarising the discussion. The advantage of structuring the outpatient clinic in this way is that the young people can practice the skills they will need in adult services in a familiar environment, additionally parents have the opportunity to gain confidence that their child can discuss their health, treatment plans or concerns with members of the MDT. Any sensitive subjects the young person wishes to discuss, such as contraception or fertility, do not have to be shared with the parents. Independent discussions with the MDT may provide the opportunity for the young person to explore subjects they may choose not to discuss with their parents present.

Nurses are an essential part of the MDT and are often a fundamental part of the process in the coordination and management of transition within teams [18]. The TKW is commonly a member of the specialist nursing team; however, this role may be occupied by more than one person with some centres reporting the specialist nurse may work in conjunction with a member of the social work or medical team to manage transition [9]. The TKW should be known to the young adult and their family and should be used as their first point of contact if they encounter any concerns while navigating the adult service [14].

TABLE 1 Commonly used educational programmes

Programme name	Ready Steady Go	CF RISE	On Trac
Country	UK and the Netherlands	USA	Canada
Key features	Resources designed to deliver a high-quality transition for young people across all healthcare subspecialties Improves clinical practice and clinical outcomes Empowers young people to manage their healthcare confidently and successfully in both paediatric and adult services	Focus on: responsibility, independence, self-care, education Increase knowledge of CF and skills young people need to independently manage their condition Knowledge assessment Responsibilities checklist Educational resources	Designed to affect changes in policy, clinical practice, communication, documentation and processes to improve transition for families and the competencies of healthcare providers Checklists Individualised sections for young adults and their parents Tips for successful transition
Further information	UK: www.readysteadygo.net [#] The Netherlands: www.opeigenbenen.nl	www.cfriase.com	www.bcchildrens.ca/our-services/support-services/transition-to-adult-care

[#]: respiratory link under construction (www.readysteadygo.net/respiratory.html).

Tools to help organise a good transition process

A shared transition protocol helps to organise the transition process. The protocol must be agreed by both the paediatric and adult CF teams and must include education programmes, the roles of the transition coordinator/TKW and team members, the transition clinics and steps to prepare young people for transfer to the adult service. All steps in a transition protocol should be used in a timeline agreed by both teams.

In preparing for transition, it is important to assess the young person's knowledge of CF and age-appropriate educational materials should be provided. These should contain information covering different aspects of CF management including lung and gastrointestinal health and treatment options. The aim of such tools is to provide robust knowledge of the disease. Some commonly used educational programmes are summarised in table 1.

Tips to prepare

Meeting the adult team

As young people and their parents learn about the transition process, it is important they meet members of the adult team. In daily practice some CF services may find this a challenge logistically; however, a meeting with the CF nurse specialist can often be arranged in person or virtually. Meeting the team at a transition clinic provides an opportunity for the young person and their family to discuss their expectations and concerns about the differences in paediatric and adult services. It is important to let the young person introduce themselves to the new team, either in person or by writing a little about themselves in the transition document. Some young people share more information than others; however, it is helpful for the adult service to gain insight into the young person's hopes for the future, hobbies and goals for future education or employment.

To help young people who transition to adult services in a different location to their paediatric care, many CF services provide short videos or tours of their facilities. These often detail the location of the hospital and introduce the service and members of the team. These resources can provide important information, such as how to get to the hospital, where the outpatient clinic is and what it looks like, and an opportunity to "meet" members of the team. A short film can be a useful alternative to an outpatient clinic before the actual transition. If CF teams do not share the same medical records system, a detailed handover letter is necessary with contact information for the paediatric CF team.

Suggested topics to include in the young person's contribution to the transition document

1. My name is:
2. The best way to reach me is:
3. Things that give me enjoyment in my life are:
4. The people I count on for support are:
5. When I am sad or am having a hard time with something, I like to (exercise, talk to friends, meditate, something else):
6. How do I feel about transferring my CF care to an adult CF team?
7. One of my goals for this year is:

8. What I like most about CF clinic or hospital stays:
9. What I like least about CF clinic or hospital stays:
10. One of the things I worry most about in life or my health is:
11. If I could change anything about my treatments it would be:
12. What topics or questions about CF do I want to learn more about this year?
13. Anything else important I would like my new adult CF team to know about me:

Challenges or risks if transition does not go well

Failure to develop skills to self-manage care

As previously mentioned, it is recommended that young people begin to practice and develop skills to manage their disease independently prior to transition. For example, by beginning to practice skills such as contacting their care team independently. The aim of this is to prepare the young person to become more autonomous in self-management and prepare them for adult services. However, this may present a challenge, as not all young people have reached a level of maturity where they are willing to start taking over responsibility for care or they may lack the confidence to do so. Many adults with chronic diseases often require support from healthcare professionals, family members or carers to enable them to successfully manage their day-to-day care [19]. GRAVELLE [20] supports this by suggesting that encouraging young people to independently manage their health when they are unwilling to do so could lead to them becoming overwhelmed and resistant to developing independence, with decreased clinic attendance and lower adherence to treatment regimens. Transition takes place at a time when young people are typically beginning to develop independence in other areas of their life, making decisions regarding their future, education and employment, and beginning relationships or moving away from home. Therefore, treatment may not be a priority, their parents may be less involved with management of treatments and adherence to treatment may decrease, this may lead to an increase in symptoms and prolonged periods of illness [21].

Young people who feel empowered to independently manage their disease report increased quality of life; therefore, it is important that young people are encouraged to begin developing self-management skills with support from their families and healthcare team gradually over time in preparation for a successful transition [22]. ZACK *et al.* [23] suggest that the ideal time to begin developing independent skills, such as speaking with a doctor alone at clinic visits, would be between 13 and 16 years of age. CRONLY and SAVAGE [19] found that encouraging young people to reflect on their adherence to treatment and its positive consequences allowed them to gain awareness of their ability to influence their health, thus acting as positive reinforcement. In turn, this increased the likelihood they would engage in healthy behaviours in the future which would positively influence their health.

Disengagement from the transition or CF service

Apprehension about any new situation is understandable, whether a new school, job or healthcare team. However, this should be addressed during the transition process and should not prevent transitioning into the adult service [24]. When transition does not run smoothly, for example if the young person or their family fail to engage in the process or if there is a no formal transition programme, there is an increased risk of young person/family disengagement and loss of follow-up in the adult service [25]. In falling between both services, the young person may not access CF-specific healthcare until the need arises, often during a deterioration in symptoms. During transition the first appointment in the adult service must be agreed by both the young person and their family, and appointments should be monitored by the TKW to ensure the young person attends clinic and is given a follow-up appointment [25]. GRAVELLE [20] suggests that paediatric and adult teams should share responsibility for follow-up of young people who fail to attend their first scheduled clinic appointment with the adult service.

Preventing gaps in care is a key focus of the CF clinical care guidelines in the USA. SAWICKI *et al.* [26] sought to identify the factors associated with disengagement at the point of transition by examining the CF Foundation patient registry. They found that young people lost to follow-up usually had not attended clinic regularly in their last year of treatment under the paediatric team. Young people were lost to follow-up for several reasons, but there was an increased likelihood of disengagement if they moved to a different city, particularly if links between paediatric and adult CF centres were absent. Furthermore, these young people were unlikely to have cultured methicillin-resistant *Staphylococcus aureus* (MRSA) and had decreased episodes of pulmonary exacerbations, suggesting that some had lost contact because they felt well and did not need to access CF services. Disengagement was also more likely if young people did not have health insurance, suggesting they came from lower socioeconomic backgrounds. Therefore, the TWK within the adult team should monitor young people who fall within these groups to ensure they are actively encouraged to engage with the adult service and are not lost to follow-up [20].

The transition process in daily practice

Challenges in daily practice

There is a wealth of research regarding the importance of a positive transition for young people with chronic diseases such as CF; however, in daily practice there are many challenges to this. A variety of factors can hinder a good transition process, with potentially major consequences for the health of the young person. Some authors suggest that transition for young people with CF is particularly challenging because it occurs at a time when there is an increased risk to health [27]. Being flexible around the timing of transition can be beneficial. On rare occasions medical instability, transplant assessment or end-stage disease may delay the process or it may not be appropriate at the time. Thankfully these circumstances are rare in modern CF management.

Organisation of the transition process can pose challenges. CF services may differ in size and small centres that transition less often have less experience in the transition process and may transition young people on a more *ad hoc* basis. Other smaller centres may cluster their services to devise a local protocol for their CF centres. However, there is little national or international consensus on how best to organise a transition programme [9].

Preparation

Preparing young people and their families for transition can be a challenge, some young people feel ready to transition whereas others may not feel as prepared. For young people transition takes place at a time of change, including education, employment and developing relationships. Therefore, transition may be the focus for the CF team, but not for the young person. Many young people transition to adult services in good health, which may also contribute to transition feeling less of a priority [12]. The young person may just want to live a normal life, with as little burden of CF as possible. Conversely, some young people may require ongoing assistance with transition, for example those with developmental delay who may require prolonged support or assistance to navigate a multitude of services. The TKW should provide prolonged assistance to those who have severe medical problems, poor understanding, or less knowledge of CF.

Paediatric care tends to be focused upon the family, whereas adult care is primarily focused upon the individual, it may take some time for young people and their families to adapt to this change. It is generally accepted that discussion about transition (including independence and knowledge of CF) should take place from at least early adolescence, although often CF teams begin discussions earlier to ensure that transition is accepted as an expected milestone in the process of growing up with CF. Beginning to educate young people about disease management early can normalise treatment, as it becomes part of the young person's daily life and they work towards becoming independent in the management of their condition.

Confidence in the expertise of the adult CF team and the differences between paediatric and adult approaches may influence the success of the transition process. Therefore, it is important for the young person and their family to meet the adult CF team as part of the transition process. Several factors could disrupt this, such as lack of time, understaffing or disinterest/disengagement from both the young person and their family.

More recently, the coronavirus disease 2019 (COVID-19) pandemic has led to changes as people with CF report increased vigilance around cross-infection issues, a reluctance to make long journeys to hospital, and a move from physical to virtual reviews or a mixture of both. It is important to consider the wishes of the young person and their family, particularly during a process of transition.

Conclusions

There are key points to be aware of when considering the transition process. During transition, healthcare must be complete, coordinated, continuous and at a period of relative stability. Countries differ regarding the age of transition; however, it is generally accepted that transition should take place around the age of 18 years [9]. It can however be earlier; for example, in the UK young people transition at 16 years, while in the USA transition can be at 21 years. A transition process must be agreed and committed to by both the paediatric and adult CF teams. A shared vision is helpful to establish a good transition process.

The transition process will be coordinated by the TKW who may be based in either the adult or paediatric team. In many CF centres the CF nurse specialist has the role of transition coordinator; however, participation from both paediatric and adult CF teams is necessary to establish an effective transition process [28].

Meeting the adult CF team is an integral part of the transition process. Research suggests that this is a vital step which helps to decrease anxiety of both young people and families, as they have the opportunity to

meet the future team and ask questions about the service and their place within it. Both parties can then discuss expectations, and the responsibilities of both the young person and the adult CF team. Practical information should be provided as well as information about the differences between the teams.

Planning the pace and process of transition should be flexible and based on the young person's capabilities and needs. Naturally the wider family will be involved in the transition process; however, ultimately the young person should be encouraged to make decisions. A complete transfer of medical and psychosocial information is essential; usually this is provided as a transition document or referral letter with a full medical and psychosocial history.

Transition does not end at physical transfer to the adult service. Support should be provided by the TKW for at least the first 6 months [3], and for some a greater level of support may be required for a longer period, for example for those with complex medical or social needs. Once they have transitioned the young person may need time to get used to the culture, a new way of working and becoming more independent and responsible for their health. Although the entire purpose of transitioning is to equip young people with the tools to become more independent in the management of their CF, ongoing support from the TKW may be necessary for a short period after official transitioning to adult services has occurred. The adult CF team must be aware of this and should be attentive to the needs of young people for the transition to be successful [17]. The importance and need for a TKW overseeing the transition process is key, as they provide continuity of care and oversight of the service from the beginning of the transition process to the young person being settled within the service.

Key points

- Transition usually takes place between 16 and 21 years of age; this varies internationally.
- Transition discussions with young people and their families should begin from at least early adolescence, but may take place sooner to normalise transition as a positive and expected process.
- Paediatric and adult CF services should work together to devise a shared transition process or protocol for patients moving between their centres.
- Meeting the adult team prior to transition allows patients and their families to ask questions about the adult service and prepare for transition, leading to decreased anxiety for both young people and their parents about the transition process.
- Teams should ensure continual engagement by CF services at the point of transition and if the young person disengages should try to re-establish contact with the young person and their family.

Self-evaluation questions

1. Is a transition protocol necessary for a positive transition process?
 - a) No, a shared vision is sufficient.
 - b) Yes, it is important for both teams that the shared vision and the transition process is documented.
 - c) No, as long as both CF teams meet regularly before the transfer.
 - d) Yes, without a transition protocol there is no positive transition process.
2. When should transition begin?
 - a) At an early age.
 - b) A year before the actual transfer.
 - c) In early adolescence.
 - d) At birth.
3. Which member of the CF team is most often placed in the role of TKW?
 - a) The physiotherapist.
 - b) The nurse.
 - c) The doctor.
 - d) The psychologist.
 - e) All of the above.
4. Is a transition document really necessary?
 - a) No, the young person can tell the doctor in the adult team her/his medical problems.
 - b) Yes, it is good for the young person to have an overview of all medical and social issues.
 - c) No, if both teams meet before the transfer all the essential information is shared.
 - d) Yes, it is beneficial for the team and the young person to have an overview of all issues and the young person can add information they think is important.

5. Which is/are the most important aspect(s) of meeting the adult CF team for the young person and their family prior to the transfer?
 - a) The young person and their family get essential information about the adult CF team.
 - b) It decreases anxiety and concern for the family.
 - c) It helps the adult CF team to know how to approach the young person and their family.
 - d) If they don't get along, the young person can choose another CF team.

Conflict of interest: D. Office has nothing to disclose. I. Heeres has received grants or contracts from Vertex, outside the submitted work.

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Suggested answers

1. b.
2. c.
3. e.
4. d.
5. a and b.