A Pilot Study Exploring the Role of Physical Therapists and Transition in Care of Pediatric Patients with Cystic Fibrosis to the Adult Setting

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ABSTRACT

Background: Cystic fibrosis (CF) is a disease that requires intensive multidisciplinary care, including care by physical therapists (PTs). People with CF are now living well into adulthood, necessitating a transfer of care from the pediatric setting to an adult one. Physical therapists play a large role in the care of the person with CF; however, there is little known about the PT role in transition of care. **Purpose:** To explore transition issues for people with CF from the perspective of PTs. **Methods:** An 18-question online survey was sent to PTs via an electronic CF listserv. Questions were derived from an analysis of transition literature in CF and other chronic childhood conditions. Physical therapists who reported treating people with CF gave their opinions on issues impacting transition from their perspective as well as their perception of patient and parent concerns. Descriptive statistics were used for data analysis. **Results:** A total of 26 PTs completed the survey. A majority, 61.5%, reported that there was a transition program at their facility, with 42.3% involving physical therapy. Common themes for patients and parents included feeling uncertain about: knowledge of the adult physician, acquiring pulmonary infections in the adult setting, and pace of the adult clinic. Physical therapists were concerned about adherence with airway clearance and exercise following transfer to the adult clinic. **Conclusions:** The role of PT in transition programs is quite varied. Physical therapists should address common concerns of their patients and families to improve the transition process and possibly impact adherence to the PT plan of care.

Key Words: transition of care, cystic fibrosis, physical therapy

INTRODUCTION AND PURPOSE

Cystic fibrosis (CF) is an autosomal recessive disease often diagnosed before the age of two. Those with CF have a mutation in the CF transmembrane conductance regulator (CFTR) that interferes with chloride transport.¹ In the lungs, the alteration in the movement of chloride and sodium results in the formation of excessive and thick mucus that impedes airflow and increases the risk for pulmonary infections.¹ It is often the pulmonary complications of CF that contribute to mortality.² Cystic fibrosis affects the exocrine glands in the lungs and throughout the body, causing issues in multiple systems/organs including the pancreas, the digestive system, and the reproductive system. The treatments for CF are aimed at maintaining pulmonary function, maximizing nutrition, preventing CF related co-morbidities (ie, diabetes, metabolic bone disease), improving quality of life, and increasing the lifespan.³⁻⁵ The disease itself and some of the medical interventions contribute to pulmonary infections, decreased pulmonary flow rates, exercise intolerance, aerobic deconditioning, strength deficits, and loss of bone mineral density.⁵ Many of these impairments can and are addressed by physical therapy through the use of airway clearance techniques (ACT), aerobic exercise, and strength training.

Approximately 1,000 children are diagnosed with CF each year with a prevalence of 30,000 children and adults in the United States living with this disease.⁶ Previously, CF was considered a pediatric disease, to be managed by a pediatric team, as most patients did not survive far into adulthood. Due to improved treatments for CF, it is now estimated that those born with the disease in the 1990s will live into their 40s, and those born in the early 2000s will live into their 50s.^{7,8} This improvement in longevity translates to the need for patients to transition from a pediatric care setting to an adult health care setting. In 1996, the CF Foundation mandated that approved CF centers providing adult CF care begin to address the need for specialized centers for adults living with CF.⁹

Transition refers to “the purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centered to adult-oriented

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health care systems.”^{9(pe1161)} Transition programs are not consistently employed throughout the United States, nor is there standardization or evidence to support a particular model. Moreover, there is no information on the role that physical therapists (PTs) play in helping their patients transition to physical therapy in the adult setting. As PTs are key members of the CF team, it is important to characterize the role that they presently play in transition. Taking stock of the present role will enable our profession to establish a plan to solidify the transition of patients to an adult PT. By better preparing pediatric patients for self-management in the adult setting, we may be able to impact adherence with the PT plan of care in the adult setting where there could be less support for the management of these important health behaviors. The purpose of the present study was to elucidate the practice of PTs as it relates to transition of care for patients with CF. Moreover, we sought to better characterize the barriers and concerns about transition from the perspective of the physical therapist.

METHODS

Licensed PTs that belong to the CF-Physical Therapy listserv administered by the Cystic Fibrosis Foundation were invited via E-mail to partake in this survey. This listserv is an on-line resource and communication tool for PTs and others interested in the care of people with CF. A link to the voluntary online survey was embedded in an E-mail invitation, and completion of the survey was interpreted as informed consent. Contact information for the authors of the study was provided so that participants could ask questions prior to completing the survey. The survey was launched through SurveyMonkey and data were collected by this service without identifying information of participants. The study was approved by the University of the Sciences Institutional Review Board. The survey questions were derived from the literature on transition of care in pediatric chronic conditions including CF.^{8-15,17,20,23,28} The PubMed database was searched with the following key terms in isolation and combination: cystic fibrosis, transition of care, transfer of care, airway clearance (AC), and adherence. Common themes from the articles identified were culled by the research team and used to craft the questionnaire.^{2,8-10,13-29} Moreover, the findings of a nationwide survey on present transition practices in CF care were used to design the survey.⁹ The PT survey consisted of 18 open- and closed-ended questions designed to determine issues surrounding transition including: present practice of physical therapists in transition programs, issues that patients and their parents raised to the PT about transition, concerns of PTs regarding transition, perceived patient adherence to ACTs before and after transition, and independence with ACT (Table 1).

RESULTS

The survey was completed by 26 PTs. A majority of the respondents reported working in acute care while the remaining worked in outpatient, academia, or “other” (Figure 1). None of the PTs that participated in this study

had a caseload that was solely devoted to people with CF (Figure 2). A majority of respondents reported that there was a transition program in place for their CF clients but less than half of the PTs indicated that physical therapy was part of the transition program.

A wide range of responses were given for the ideal time to initiate transition discussions and processes. Most of the respondents indicated that transition discussions should occur between 12 to 18 years old (Figure 3). When asked what content PTs should focus on to improve transition, responses included: increasing understanding of the importance of exercise and ACT, increasing independence with exercise and ACT, importance of posture, issues surrounding urinary incontinence, monitoring and progressing physical activity, and the role of the adult CF PT. Respondents also noted that meeting the adult CF PT before transfer was important.

In terms of estimating future adherence of their pediatric patients after they transition to the adult setting, participants reported that they lost many patients to follow-up. Those that could follow their patients into the adult setting estimated that their patients adhered to the PT plan of care a majority of the time; however, there was quite a bit of variation in their estimations of adherence and their level of independence with ACT (Figures 4, 5). The respondents listed a variety of ACT used by their patients with a majority using High Frequency Chest Wall Oscillation, followed by oscillatory positive expiratory pressure devices (PEP) and standard PEP (Figure 6).

The PT respondents perceived their patients with CF and their parents had the following concerns about transition: not having enough insurance coverage in the adult setting, not receiving the same quality of care in the adult setting, lack of knowledge about CF in adult based clinic, quicker pace in adult setting, risk of acquiring pulmonary infections, and timing of the transition while other major changes are happening (Figure 7). There was also a concern about getting closer to the end of their expected lifespan. The most common concern patients reportedly expressed to their PT was fear of receiving lower quality of care in the adult setting. The most commonly reported concern that parents expressed to the PT was the timing of transfer, as it occurs when children are going to college. The PTs themselves reported that their most common concerns surrounding transition and transfer to the adult setting were: drop in adherence with exercise, an increase in risky behaviors such as drinking and promiscuity, and a drop in adherence with ACT.

DISCUSSION

The purpose of this study was to explore issues surrounding the transition of children with CF from a pediatric setting to an adult setting from the perspective of PTs. The total number of PT respondents was 26, with the majority working in an acute care setting. All of the respondents were responsible for a mixed case load, with the majority having less than half of their caseload focused on people with CF. Most reported that there was a

Table 1. Online Survey

In this survey we are asking about the transition of children with cystic fibrosis from the pediatric setting to the adult setting. Transition refers to children taking on more responsibility as it relates to the care of their body including; making and attending medical visits, following through with airway clearance and exercise, and making decisions in partnership with the medical team about their medical care.

1. What age group do you primarily treat in pediatric physical therapy?
 - a. Outpatient
 - b. Acute Care
 - c. Post-acute care - (acute rehab, sub-acute rehab, SNF)
 - d. Long term care facility
 - e. I am primarily involved with education/academics
 - f. I am primarily an administrator of a Physical Therapy clinic
 - g. Other _____
2. How much of your daily case load is dedicated to treating children with cystic fibrosis?
 - a. 100%
 - b. 75-99%
 - c. 50-74%
 - d. < 50%
3. Does your facility have a specific transition program in place to help patients with cystic fibrosis move from pediatric to adult care facilities?
 - a. Yes
 - b. No
4. Is a physical therapist part of the transition program?
 - a. We don't have a specific transition program
 - b. No, physical therapy is not involved in the transition program
 - c. Yes, physical therapy is involved in the transition program
5. At what age do you believe children should START the transition process from the pediatric setting to the adult care facility? This may include things like seeing the physical therapist or physician without parents, beginning to be responsible for their airway clearance program, etc.

FREE TEXT RESPONSE
6. What role do you think physical therapy should have in the transition of people with CF from the pediatric to the adult setting?

FREE TEXT RESPONSE
7. What topics should a physical therapist cover to prepare their patients for a transition to adult medical care?

FREE TEXT RESPONSE
8. How important do you think it is for the patient to meet the adult Physical Therapist prior to transitioning?
 - a. Not important
 - b. Somewhat important
 - c. Neutral
 - d. Important
9. In your experience, what concerns do your pediatric patients raise regarding a transition to an adult setting?
 - a. Not having enough insurance coverage in the adult care facility
 - b. Not receiving the same quality of care in the adult setting as received in the pediatric facility
 - c. Lack of knowledge about CF in adult based physician
 - d. Quicker pace in adult setting medical visits
 - e. Infections in the adult setting
 - f. Transitioning to the adult setting means that they are closer to the end of their life
 - g. Other (TEXT BOX)

10. In your experience, what concerns do the parents of your pediatric patients raise regarding a transition to an adult setting?
 - a. Not having enough insurance coverage in the adult care facility
 - b. Not receiving the same quality of care in the adult setting as received in the pediatric facility
 - c. Lack of knowledge about CF in adult based physician
 - d. Quicker pace in adult setting medical visits
 - e. Infections in the adult setting
 - f. Transitioning to the adult setting means the person is closer to their end of life
 - g. Transitioning during a time where other big transitions in life are occurring such as college
 - h. Hearing information regarding their CF care or condition since their caregivers are no longer shielding them from information in the adult care setting
 - i. Other (TEXT BOX)

11. In your experience, what are some common concerns about transition from the physical therapist perspective? (CHECK ALL THAT APPLY)
 - a. Lack of frequent visits to the CF team in the adult setting
 - b. Adult physical therapist don't have a bond with the patients
 - c. Adult physical therapist knowledge about CF
 - d. Drop in compliance with airway clearance
 - e. Drop in compliance with exercise
 - f. Increase in risky behaviors (drinking, sexual promiscuity, etc.)
 - g. Lack of preparation for the adult setting
 - h. Transitioning during a time where other big transitions in life are occurring such as college
 - i. Hearing information regarding their CF care or condition since their caregivers are no longer shielding them from information in the adult care setting
 - j. OTHER (FREE TEXT)

12. In your experience, how would you estimate the compliance that your pediatric patients have with an aerobic exercise program once they move into the adult setting?
 - a. I don't know, as I lose these patients to follow up
 - b. Compliant less than 25% of the time
 - c. Compliant 25-50% of the time
 - d. Compliant 50-75% of the time
 - e. Compliant >75% of time

13. In your experience, how would you estimate the compliance that your pediatric patients have with a strength training exercise program once they move into the adult setting?
 - a. I don't know, as I lose these patients to follow up
 - b. Compliant less than 25% of the time
 - c. Compliant 25-50% of the time
 - d. Compliant 50-75% of the time
 - e. Compliant >75% of time

14. In your experience, how would you estimate the compliance that your pediatric patients have with their airway clearance program once they move into the adult setting?
 - a. I don't know, as I lose these patients to follow up
 - b. Compliant less than 25% of the time
 - c. Compliant 25-50% of the time
 - d. Compliant 50-75% of the time
 - e. Compliant >75% of time

15. At the time of transition, how much assistance do your pediatric patients need with their airway clearance techniques?
 - a. They are independent with airway clearance
 - b. They need assistance less than 25% of the time
 - c. They need assistance 25-50% of the time
 - d. They need assistance 50-75% of the time
 - e. They need assistance 100% of time
 - f. Patients don't transition to an adult setting

16. Which of the following airway clearance modalities do you prescribe for the majority of your patients?
 - a. PEP with vibration (ie, Acapella)
 - b. PEP mask
 - c. Chest physical therapy
 - d. High frequency chest wall oscillation (ie. The Vest)
 - e. Other (FREE TEXT)

We appreciate the time you've taken to complete this survey. We welcome additional comments regarding the role of the physical therapist in transition of care for people with cystic fibrosis.

OPEN TEXT

Figure 1. Physical therapist practice setting.

Figure 2. Caseload devoted to cystic fibrosis.

Figure 3. Recommended age to initiate transition (n=20).

transition program in place; however, less than half stated that physical therapy was a part of the program.

Transition refers to the process of preparing the child/adolescent psychologically, physically, and emotionally to be more independent and directive in their care in preparation for entering an adult health care system. The transition process should span a few years to enable the child and their family to prepare.⁹⁻¹² Pre-established programs in diabetes, sickle cell, congenital heart disease,

and juvenile rheumatoid arthritis use different time frames to initiate transition with some starting as early as 11 to 12 years old,¹³ and some between the ages of 13 and 16.^{14,15} Other studies indicated that transition should begin as late as 18 or 21 years of age,¹⁶ underscoring the lack of agreement on when this process should begin. The lack of consensus about transition was also seen in this current study. Among respondents to the survey, there was a clustering of responses indicating that many believe transition should occur between 12 and 18. In a survey of CF programs in the United States, the median age for transition was 17, with transfer occurring around age 19.⁹ Only 28% of those facilities offered visits that focused on transition.⁹

Many transition programs for children with chronic diseases lack a uniform process or consistent written guidelines for what content to include in the program.^{9,13,17-23} In this current study, respondents indicated that PTs should include information on the importance of exercise in managing CF, monitoring and progressing exercise, the role of exercise in mediating loss of bone density, exercises for urinary incontinence, and the importance of airway clearance. Moreover, the majority felt that meeting the adult-based PT prior to transfer into the adult setting was an important part of the process.

Transition and transfer into a new health care setting is often an uncertain time for adolescents and their parents.¹¹ This current study indicated that PTs perceived both patients and parents to be concerned about insurance coverage (as dependent care often ends at age 21), CF knowledge of the adult physician, fast pace in adult clinics, risk of infections, and transferring when also beginning college. Concerns about quality of care and the ability of children to take care of themselves have been raised by other authors as additional issues surrounding transfer of care.^{11,13,15,16,18,20-22,24-27}

Previous researchers have found that children with CF and their parents have concerns about infection acquisition in the adult setting.^{8-11,13,14,17,18,21,24} Parents have also expressed concern about overloading the patient with information about their prognosis, especially if the parents had shielded their child from this information previously.^{11,13,15,16,18,20-22,24-27} Parental concerns noted in previous research included (1) the realization that their child transitioning to adult care meant the child is closer to the end of life^{24,28} and (2) the difficulty parents had of letting go of the primary responsibility of the child's health care after being involved for years.^{17,21,22,24,28,29} In our current study, PTs were mostly concerned about adherence with the physical therapy plan of care (exercise and ACT), as well as the increased risk for behaviors such as drinking, drugs, and promiscuity.

Transition and transfer also bring about concerns of adherence with care, including exercise and ACT. Our study revealed that PTs estimated their patients' adherence with aerobic exercise, strength training, and ACT were less than ideal when they transferred to the adult setting. Studies in other populations show mixed results, with one study by Tuchman et al¹⁷ revealing that patients after

Figure 4. Estimated adherence with physical therapy plan of care in the adult setting.

Figure 5. Amount of assistance needed with airway clearance techniques at time of transfer.

Figure 6. Type of airway clearance reportedly used by patients.

liver transplant or with congenital heart disease did have lowered adherence to their medical regimens and poorer health status after transfer. In CF, a study by Dugueperoux et al²⁹ did not show any difference in the rate of decline in pulmonary function tests when comparing them one year before transfer and one year after. Moreover, the occurrence of common comorbidities such as diabetes

Figure 7. Concerns about transition and transfer to adult cystic fibrosis centers as perceived by physical therapists.

wasn't significantly different one year after transfer, nor was the rate of infections.²⁰ Less is known about the impact that transfer has on adherence rates for ACT and exercise, although the participants in this current study postulate that adherence does in fact drop off.^{2,3}

There are many limitations to consider when interpreting the results of this pilot investigation into the role of the PT in transitioning patients with CF to an adult care setting. The primary limitation is that the information regarding patient and parent concerns about transition was not acquired directly; rather it was through the lens of the PT. This introduces bias, as the PT is making inferences that may not be valid. Another area for consideration is response bias. Detailed information regarding the respondents practice, geographic location, or educational background was not collected, limiting the ability to characterize those that participated in this survey. Other limitations include the small sample size and potential ambiguity regarding the definitions for transition and transfer of care.

CONCLUSION

There is a lack of consistency in the timing and content of transition programs for people with CF. As life expectancy is increasing, there is a need to help children with CF become healthy independent adults with CF through a coordinated and evidence-based transition program. As PTs play a large role in the care of people with CF, it is imperative that the profession actively seeks to be a part of transition programs and create appropriate transition goals for ACT and exercise.

In this study, PTs did not consistently take part in transition programs for children with CF. There is a need for a more unified, evidence-based and developmentally appropriate approach to transitioning people with CF to adult based physical therapy. In order to reach this goal, additional research is necessary to determine the key components of a physical therapy transition program, with information gained directly from patients and their parents. Physical therapists have a unique opportunity to solidify their role in transition programs to help foster lifelong

independence, possibly improving the quality of life for adults with CF.

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