Improving Health Service Access for Youth with Cystic Fibrosis: A Narrative Review

By: Keri Durocher[1], Dr. Jamie Crawley[2] & Dr. Jody Ralph[3]

[1]This paper was originally conceptualized and written for the course, *The Health of Individuals, Families and Groups* in April 2019, which is a component of the Master of Nursing program at The University of Windsor.

[2]Dr. Jamie Crawley and [3]Dr. Jody Ralph are Associate Professors at The University of Windsor. Both authors made substantial contributions to the revision and editing processes of the manuscript.

Corresponding author: Keri Durocher, PhD Student- Arthur Labatt Family School of Nursing, Western University, London, Ontario, N6A3K7; kduroch3@uwo.ca

Abstract

Background and Purpose: Cystic Fibrosis (CF) is a complex condition that impacts various functions throughout the body. Improvements in health through technological advances and research have increased the global life expectancy for youth with CF; however, access to resources and support remains suboptimal. Therefore, our primary aim was to identify the most common access issues for youth with CF, with secondary aims of raising awareness about these barriers and proposing potential solutions.

Methods: An initial literature scan was conducted, where five key themes were identified as the most pertinent access issues for youth, including: treatment complexity, distance to care facilities, financial constraints, family involvement, and mental health factors. To explore these access issues in further detail, we conducted an iterative narrative review across four databases.

Results: Twenty-five research articles were identified across the five themes, and included the following designs: review, cross-sectional, survey/questionnaire, descriptive, quasi-experimental, longitudinal, cohort, observational, and qualitative research. Expert opinions and news articles were also included using a systematic grey literature scan.

Conclusion: While these articles provide the most up-to-date evidence on access issues for youth with CF, current research remains limited. However, nurses can be key advocates for assisting youth populations with CF to better access health services while minimizing potential barriers to care. Some proposed advocacy activities include supporting policy changes through funding for life-enhancing medication and increasing access to digital health services that improve health outcomes for youth with CF.

Keywords: Cystic Fibrosis; access; healthcare; health services; youth; digital health

Introduction

Access to necessary health services for youth with Cystic Fibrosis (CF) is a critical and pertinent issue, especially due to the various factors hindering accessibility that are more prevalent in adolescent years. Treatment adherence throughout youth is usually sub-optimal; however, the diverse reasons for this problem have not been well-addressed (Faint et al., 2017). According to Faint et al. (2017), some growth and development factors that may impact adherence for youth include the desire for social acceptance and multiple body changes that occur during the adolescent years. However, treatment adherence during youth is crucial to longevity, as performing excellent self-care provides a foundation for ongoing adult health and well-being. Furthermore, the risk of developing lifelong complications as a result of the disease, including liver complications and Cystic Fibrosis-Related Diabetes (CFRD) is greater during this time period (Cystic Fibrosis Canada, 2020). Access issues are complex factors that may impact treatment adherence for youth, and therefore, it is necessary to identify the issues specific to this population and to propose possible solutions. Nurses play a crucial role in assisting with key care issues and can advocate for policy changes. The overall goal of exploring these topics is to enhance the quality of life for youth with CF to help them thrive in adulthood.

Background: Cystic Fibrosis Epidemiology, Pathophysiology and Current Treatments

Cystic Fibrosis (CF) is an autosomal recessive disorder that impacts 70,000-100,000 people worldwide (Graham & Hart, 2021; Shteinberg et al., 2021). In 2018, the median life expectancy for people with the most common CF mutation was over 43 years of age for those living in the United Kingdom, but under 15 years for those living in less developed countries (Graham & Hart, 2021). This statistic highlights the positive impact that new technologies for diagnosis and treatment regimes have on life expectancy. The shift in epidemiology from pediatric to adult CF populations also creates new challenges that the healthcare system should adapt to (Shteinberg et al., 2021).

Cystic Fibrosis is caused by more than 2000 mutations in the cystic fibrosis transmembrane regulator (CFTR) gene. CFTR encodes for a cAMP-regulated chloride channel located in the membrane of secretory epithelial cells of sweat glands, respiratory airways, the gastrointestinal tract, pancreas, and vas deferens (Maule et al., 2020; Shteinberg et al., 2021). The mutated CFTR gene alters salt and fluid regulation, resulting in thick mucus secretions, reduced mucociliary clearance, chronic pulmonary infection and inflammation, and eventual multiorgan dysfunctions and respiratory failure (Maule et al., 2020; Shteinberg et al., 2021). Disease severity is thought to have a genetic influence in terms of mutation class, modifier genes, and variable alleles, which may also determine responsiveness to new CFTR modulators (Shteinberg et al., 2021). The six classes of CFTR mutations correspond to defects in the quantity and/or production of CFTR mRNA or protein products. For example, Class I mutations result in a complete absence of protein production and Class V mutations result in a reduced protein production (Cutting, 2015; Graham & Hart, 2021; Lopes-Pacheco, 2020; Shteinberg et al., 2021).

In the past, treatment strategies for CF were limited to symptom management, rather than addressing the altered CFTR gene (Maule, Arosio, & Cereseto, 2020). Even now, the most time-consuming, frequent, and often physically uncomfortable treatments focus on clearing the airway of mucus; treatment strategies such as these consume two hours a day on average (Graham & Hart, 2021). For some individuals, CFTR modulators have demonstrated significant efficacy. CFTR modulators are small molecules that improve CFTR protein function (Maule, Arosio, & Cereseto, 2020; Pinto, Silva, Figueira, Amaral, & Lopes-Pacheco, 2021; Shteinberg, Haq, Polineni, & Davies, 2021). High-throughput assays are used to screen for and identify the effective CFTR modulator therapies (Pinto et al., 2021). Unfortunately, more than 10% of patients cannot benefit from CFTR modulators, as they have rare mutations or CFTR mutations that are not responsive to CFTR modulators (Lopes-Pacheco, 2020; Maule et al., 2020; Pinto et al., 2021). CRISPR (Clustered Regularly Interspaced Short Palindromic Repeats) and CRISPR-Cas9 are powerful gene-editing biotechnology tools that may offer the opportunity to correct the underlying cause of CF (Graham & Hart, 2021; Maule et al., 2020). Currently, although CRISPR-Cas9 offers significant promise as a tool to "cure" CF, a significant barrier to its utilization in the clinical setting is the fact that the airway epithelium is made up of non-dividing cells where precise editing via homology-directed repair does not occur (Graham & Hart, 2021).

Methods

The primary aim of conducting this narrative review was to explore the question, "*What are the main access issues that youth with CF encounter*?" Secondary aims included the identification of barriers to raise awareness surrounding these accessibility issues, and to propose potential solutions to address these barriers.

The search strategy included searching the databases PubMed, ProQuest, CINAHL, and Google Scholar for relevant literature related to access issues. An initial scan of the literature was performed to identify the most pertinent access issues and grouping these by themes. These themes were analyzed by the primary author, who has five years of clinical experience working with youth populations with CF. Twenty-five articles were then selected for inclusion through an iterative search method focusing on the following five themes: treatment complexity, access to care facilities, family involvement, mental health support, and financial concerns. The research designs of the included articles were review, cross-sectional, survey/questionnaire, descriptive, quasi-experimental, longitudinal, cohort, observational, and qualitative studies. Articles were excluded if they did not relate specifically to youth, or if they did not centrally focus on CF. A grey literature scan was also conducted using the Google search engine to supplement the academic results. The search involved scanning both the regular results and news sections for relevant access issues as well as potential nursing implications for this topic. Supplemental literature was also included as needed, such as textbook chapters. Literature was included using all research designs, including reviews, quantitative, and qualitative studies. Year and country limitations were not set due to the novel concepts that were analyzed and the limited amount of research that has been performed to explore this issue. The articles that were selected had to be

available in the English language. A full summary of relevant topics, search terms, and included articles can be found in Table 1.

Literature Review

When planning care for youth with CF, the complex, interrelating issues that influence access to health services is imperative for nurses to understand. It is essential to note the complexity of CF care for adolescents because of growth and development, and the multi-organ involvement of this condition (Gould & Dyer, 2011). These considerations may require advocacy efforts for more effective care provision, when compared to other genetic conditions. Some of the following considerations can be well-supported by nurses and can also involve the integration of an interdisciplinary team to foster a holistic healthcare approach. Larger health system issues may also warrant political lobbying to fund health initiatives that will enhance access to treatment for this population.

Treatment Complexity

The day-to-day activities for youth with CF seem even more difficult when the busy lifestyle of someone in this age group is considered. A person with CF must adhere to a vigorous schedule, which includes chest physiotherapy two to three times per day, multiple oral and nebulized medications, ensuring optimal caloric nutrition is met, exercise, and possible care of a gastrostomy tube for extra nutrition (Cystic Fibrosis Canada, 2013; de Deus et al., 2019; Gjengedal et al., 2003; White et al., 2009). Adherence rates to treatments for younger populations is less than 50%, with higher rates for less time-consuming interventions, such as taking enzymes and oral antibiotics (Dempster et al., 2018; Goodfellow et al., 2015; White et al., 2009). It is imperative that low adherence rates are considered within the context of multiple demands for youth, which may include attending school, homework, extracurricular activities, and having a part-time job. It is important to consider that adherence rates may be negatively impacted if youth do not have access to the devices needed for these different activities. Furthermore, access to community facilities that can facilitate areas of care, such as recreational centres for exercise, can also be negatively impacted by location barriers (Denford et al., 2020). At a time when youth must take an active role in their care to maintain optimal health, managing the normal stressors of day-to-day life may overpower the desire to complete essential treatments.

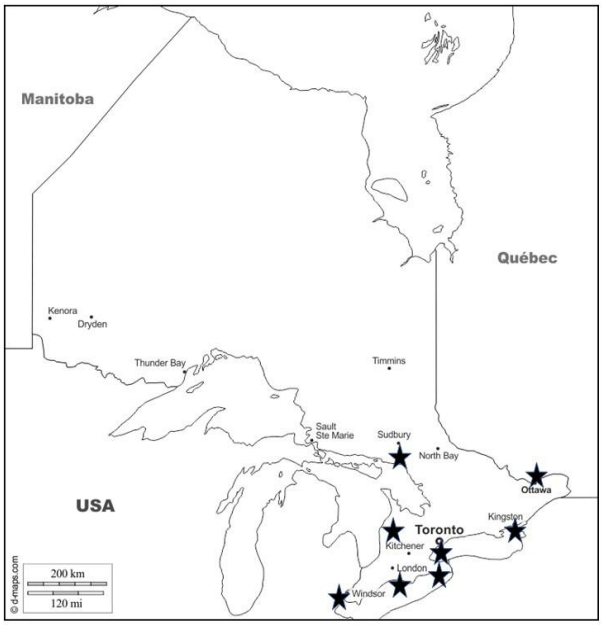
Access to Care Facilities

Attending appointments at designated care facilities is an imperative, proactive health measure as part of an overall plan of care (Cystic Fibrosis Foundation, 2020). However, it is not always easy for youth to access these clinics if they live in remote areas of their state or province (Perrin et al., 2014). For example, in Ontario, Canada, there are eight cities with continuing care clinics (Cystic Fibrosis Canada, 2020). These locations mostly spread across the Southwestern and Eastern areas of the province. However, based on 2016 census data, over 700 000 people

reside in Northern Ontario (Statistics Canada, 2016). One large city within Northern Ontario is Thunder Bay, with a population of approximately 108 000 people. It is an eleven-hour drive from Thunder Bay to Sudbury, which is the closest follow-up clinic for individuals who reside in this city (Statistics Canada, 2016). Even more of Ontario's population spreads farther north of Thunder Bay, which makes travelling to a specialized clinic practically inaccessible. Therefore, although there are CF support groups that exist in Northern Ontario, access to specialized care from CF healthcare professionals is limited (Cystic Fibrosis Canada, 2020).

Figure 1

Cystic Fibrosis Clinics in Ontario



(d-maps, 2020)

This inequitable access to specialty care based on geography is not unique to Ontario, Canada. Many of the states in the United States have one to three CF Care Centres (Cystic Fibrosis Foundation, 2020). Each state and territory of Australia has zero to two Children's CF Clinics located in the major cities (Cystic Fibrosis Australia, 2021).

Geake et al. (2020) followed the long-term outcomes of a group of children diagnosed with CF in Australia. They found that, during the 10-year follow up, many of the families moved from more remote and rural areas to regions with specialty CF care. They also found that the rate of lung transplantation was higher, and the mortality rate was lower for patients receiving centralized specialty care versus non-central care (Geake et al., 2020).

Many regions incorporate telehealth in their models of care, an initiative that increased during the COVID-19 pandemic. Prior to the COVID-19 pandemic, Cox et al. (2012) conducted a systematic review of telehealth in CF and found that participant non-compliance with data reporting was high (43-63%). Furthermore, a study that examined telehealth services within a specialty CF clinic demonstrated how 34% of follow-up phone calls were unanswered by parents (Parker-McGill et al., 2016). However, several authors reported that the COVID-19 pandemic resulted in reduced access to in-person care and increased utilization of telehealth (Colombo et al., 2020; Compton et al., 2020; Kouri et al., 2020).

Family Involvement

Due to the impressionable age of the youth population, they may be more at risk for non-adherence to treatment without the social support of their family members, or when family members are not available to assist with treatments (Bregnballe et al., 2017). A transition to independence must be balanced with the support that youth still require from their parents to avoid undue stress. In an exploratory study, effective family dynamics in the home were found to directly relate to enhanced pancreatic enzyme functioning and adherence to antibiotic treatment (Everhart et al., 2014). These positive outcomes provide preliminary data on the importance of familial support in the care of youth with CF. Conversely, this team of researchers discovered through interviews that when loved ones appear too task-oriented during day-to-day treatments, negative emotional repercussions may result (Everhart et al., 2014). Although this may not be the intention of family members, the complexity of treatment regimes may cause this perception for youth. Specific mechanisms by parents are inherently needed to avoid negative emotional responses by youth, such as practicing active coping (Zubrzycka, 2018). Effective family functioning was also studied by a team of researchers who developed a cohesion scale to assess how family functioning relates to adherence to care for youth with CF. Their findings demonstrate that high adherence to treatment correlates with families that scored high on the cohesion scale (White et al., 2009). Furthermore, promoting the accessibility of coaching family members on how to support youth is crucial to help impact treatment adherence and family relationships.

Mental Health Support

The need for mental health support for youth is imperative, as negative psychological outcomes can impact adherence to therapy and quality of life (Besier & Goldbeck, 2012). Therefore, mental health support for youth with CF is a consideration that nurses can adapt into their plan of care for this population. Youth with complex care needs require mental health interventions that encompass a multidisciplinary approach (Woodgate et al., 2018). For example, in a study on a chaplaincy intervention for individuals with CF, psychological exploration of feelings has shown to be effective (Harrop, 2007). This finding is important, as it demonstrates that social consequences of having CF have a significantly negative impact on youth when compared to other age groups, such as decreased family functioning and impaired sibling relationships, lowered school performance, and an increased risk of depression (Harrop, 2007). A Theory of Reasoned Action model was implemented in a more recent study, which discovered

that if youth feel that they have control over their treatment, better health outcomes will result (Purcell et al., 2015). This model focuses on various factors that may impact a youth's coping mechanisms, including severity of disease, familial relationships, and spiritual qualities (Purcell et al., 2015). Therefore, this model provides a framework for nurses to assess how youth may cope with the mental impact of CF based on comprehensive background information, such as demographic criteria, the severity of CF, and family functioning.

Spirituality is another concept that has been explored to assess the mental health status of youth with CF. A phenomenon called positive spirituality is a concept that youth can experience based on pre-set criteria, and this may lead to more effective airway clearance than negative spirituality (Grossoehme et al., 2016). Positive spirituality is described as finding meaningful connections in life, as well as developing healthy coping mechanisms for managing CF (Grossoehme et al., 2016). As a result, the importance of mental health awareness for this population is imperative for their overall wellbeing.

Financial Issues

Although advances in treatment have significantly improved the quality of life for youth with CF, incurred medical expenses place a large burden on families (Shardonofsky et al., 2019). As many treatments are time-intensive, these financial impacts cause a ripple effect for those who may already be economically disadvantaged. For example, a study on family system responses identified how single parents are more likely to experience perceptions of low financial well-being than two-parent families (Shepard, 1992). Conversely, a study on the lived experiences of fathers of children with CF demonstrated how even in two-parent families, 30% of fathers reported employment strain due to CF-related care needs (Shardonofsky, 2019).

There are also many hidden costs for youth with CF that may cause considerable financial strain, resulting in decreased financial wellbeing among families (O'Haver et al., 2010). Without considering additional hospitalizations, most patients with CF have quarterly appointments at their designated follow-up clinic (Cystic Fibrosis Foundation, 2020). Care for youth with CF may become a challenge when incorporating travel costs, including gasoline or airfare. When youth with CF become hospitalized, the average length of stay is approximately two weeks (Cystic Fibrosis Canada, 2020). Uninsured costs such as accommodation, food, and other living expenses for caregivers may go unreported when accounting for costs of care. In a European cost-of-illness analysis, the peak age of maximal CF-related expenses was 16 years old. With increasing complexity of treatment, this finding demonstrates the financial burden that youth with CF encounter (Mlčoch et al., 2017).

Table 1

Content Themes of Selected Articles and Associated Search Terms for Academic Review

Content Theme	Associated Search Terms	Relevant References (in order of appearance)
Treatment Complexity	Youth OR Adolesc* OR Teen* AND Cystic Fibrosis AND Treatment; Youth OR Adolesc* OR Teen* AND Cystic Fibrosis AND Med*	(Cystic Fibrosis Canada, 2013), (de Deus et al., 2019), (Gjengedal et al., 2003), (White et al., 2009), (Dempster et al., 2018), (Goodfellow et al., 2015), (Denford et al., 2020)
Access to Care Facilities	Youth OR Adolesc* OR Teen* AND Cystic Fibrosis AND Care; Youth OR Adolesc* OR Teen* AND Cystic Fibrosis AND Hospital; Youth OR Adolesc* OR Teen* AND Cystic Fibrosis AND Facility; Cystic Fibrosis AND Pandemic AND Access	(Cystic Fibrosis Foundation, 2020), (Perrin et al., 2014), (Cystic Fibrosis Canada, 2020), (Statistics Canada, 2016), (Cystic Fibrosis Australia, 2021), (Geake et al., 2020), (Cox et al., 2012), (Parker-McGill et al., 2016), (Colombo et al., 2020), (Compton et al., 2020), (Kouri et al., 2020)
Family Involvement	Youth OR Adolesc* OR Teen* AND Cystic Fibrosis AND Family; Youth OR Adolesc* OR Teen* AND Cystic Fibrosis AND Caregiver	(Bregnballe et al., 2017), (Everhart et al., 2014), (Zubrzycka, 2018), (White et al., 2009)
Mental Health Support	Youth OR Adolesc* OR Teen* AND Cystic Fibrosis AND Mental Health; Youth OR Adolesc* OR Teen* AND Cystic Fibrosis AND Psych*	(Besier & Goldbeck, 2012), (Woodgate et al., 2018), (Harrop, 2007), (Purcell et al., 2015), (Grossoehme et al., 2016)
Financial Issues	Youth OR Adolesc* OR Teen* AND Cystic Fibrosis AND Financ*; Youth OR Adolesc* OR Teen* AND Cystic Fibrosis AND Cost	(Shardonofsky et al., 2019), (Shepard, 1992), (O'Haver et al., 2010), (Cystic Fibrosis Foundation, 2020), (Cystic Fibrosis Canada, 2020), (Mlčoch et al., 2017)

Nursing Implications and Future Directions

Advocacy for Medication Availability

One way to simultaneously address treatment complexity access and financial issues is through enhanced medication availability for youth. Multiple medications developed in recent years appear to be modern breakthroughs in the care of individuals with CF. Two examples of medications that have been developed for symptomatic management of CF include ORKAMBI and Trikafta. ORKAMBI's mechanism of action leads to a decrease in viscous mucous production, which would normally be present because of ionic imbalances. The reduction in mucous leads to an improvement in overall affected organ function (Orkambi, 2019). Trikafta leads to an increase in predicted forced expiratory volume (ppFEV1), which also relates to improved lung function (Cooke, 2020). Unfortunately, neither of these medications are cures for CF, but they may result in greater alleviation of symptoms and enhanced quality of life (Cystic Fibrosis Canada, 2020).

Trikafta is currently approved for use in Canada and the United States (Cystic Fibrosis Canada, 2021; U.S. Food & Drug Administration, 2019). ORKAMBI is currently approved for use in over ten developed nations (Boisvert, 2018). However, the costs associated with obtaining these drugs are high, as the annual cost of ORKAMBI and Trikafta are approximately $250 000 and $311 000, respectively (Biopharma Dive, 2019; Braun, 2018). The inclusion criteria to be eligible for these medications also presents significant barriers. For example, in Canada, to be eligible to receive coverage for ORKAMBI, the individual must have a 20% decrease in their lung function (ppFEV1) over six months, despite effective care measures (Cystic Fibrosis Canada, 2020). This restriction may make ORKAMBI and other life-enhancing medications inaccessible to those who could benefit from their use.

According to Graham & Heart (2021), 76% of patients with CF have reported issues in acquiring necessary medications for treatment. Therefore, nurses and other multidisciplinary team members must be key advocates to publicize needed funding for life-enhancing medications, as they provide direct care to individuals with CF (Cystic Fibrosis Canada, 2020). Fostering a multidisciplinary approach will make greater strides towards federal funding of life-saving medications regardless of individual insurance status. For example, nurse researchers can undertake preliminary studies to find empirical evidence that supports the use of these medications. This experimental research is vital to support nursing evidence that youth need these medications to improve their well-being.

Digital Technology Support

Distance to care facilities, enhancing mental health support, and improving the availability of family member involvement may be addressed through digital health solutions. The COVID-19 pandemic has highlighted the necessity of having digital access support for youth with CF. Although telehealth services were not routinely utilized prior to the pandemic, a recent increased uptake in digital services shows how they can be a viable solution to enhance care for youth with CF in the future, including home monitoring for pulmonary function, adherence to treatments, exercise, and self-management (Cox et al., 2012; Calthorpe et al.,

2020). Nurses must be diligent in promoting these services to youth at the point of care, which can involve explaining the benefits of digital health and advocating for the infrastructure that is needed to support youth in making transitions to digital health care.

Conclusion

After describing the many reasons why access to health services for youth with CF may be increasingly complex, it is crucial to acknowledge that care for this population cannot be ignored. All too often, youth are categorized as non-compliant with treatment; however, this is too simplistic of a conclusion used to explain why they may not be actively engaged in health-sustaining measures. After reviewing the barriers experienced by youth with CF, it is imperative that action is taken to implement policy changes that will make a lasting positive impact. Specifically, nurses should consider key accessibility issues, including treatment complexity, access to care facilities, family involvement, mental health support, and financial issues. Lastly, nurses can be effective advocates and experts for this population through solutions such as advocating for medication availability and enhancing digital technology support. If action is taken to further improve health service access for youth with CF, health outcomes for youth can continue to improve in the future.

Declaration of Interest Statement: The authors declare no conflict of interest.

Funding Sources: None

Acknowledgements: We would like to thank Anh (Joanne) Ta and Rachelle Charon for their assistance in this work.

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