Improving chronic care delivery and outcomes: the impact of the cystic fibrosis Care Center Network

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ABSTRACT

Cystic fibrosis (CF) is a multisystem, life-shortening genetic disease that requires complex care. To facilitate this expert, multidisciplinary care, the CF Foundation established a Care Center Network and accredited the first care centres in 1961. This model of care brings together physicians and specialists from other disciplines to provide care, facilitate basic and clinical research, and educate the next generation of providers. Although the Care Center Network has been invaluable in achieving substantial gains in survival and quality of life, additional opportunities for improvements in CF care exist. In 1999, analysis of data from the CF Foundation's Patient Registry detected variation in care practices and outcomes across centres, identifying opportunities for improvement. In 2002, the CF Foundation launched a comprehensive quality improvement (QI) initiative to enhance care by assembling national experts to develop a strategic plan to disseminate QI training and processes throughout the Care Center Network. The QI strategies included developing leadership (nationally and within each care centre), identifying best CF care practices, and incorporating people with CF and their families into improvement efforts. The goal was to improve the care for every person with CF in the USA. Multiple tactics were undertaken to implement the strategic plan and disseminate QI training and tools throughout the Care Center Network. In addition, strategies to foster collaboration between care centre staff and individuals with CF and their families became a cornerstone of QI efforts. Today it is clear that the application of QI principles within the CF Care Center Network has improved adherence to clinical guidelines and achievement of important health outcomes.

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INTRODUCTION

Cystic fibrosis (CF) is a complex, multisystem, genetic disease caused by abnormalities in the CF transmembrane conductance regulator (CFTR), which is a chloride transporter that resides on the surface of specialised epithelial cells. Significant advances in genetic and biomedical research have enhanced understanding of the disease, its cause, clinical management and approaches to diagnosis. Although still without a cure, CF is no longer considered a terminal childhood disease, but it remains a life-limiting condition with a median life expectancy of 41.1 years in 2012[1] (figure 1). Currently, almost 50% of the individuals with CF in the USA are adults (figure 2).[1] Advances in care delivery have the potential to extend survival and improve the quality of life of affected individuals.

CLINICAL MANIFESTATIONS OF CF

The clinical manifestations of CF are due to CFTR dysfunction in the affected organs including lungs, pancreas, bowel, vas deferens, sinuses and liver. The hallmark of CF is progressive, obstructive lung disease due to abnormal airway secretions, leading to chronic airway infection, excessive inflammation, bronchiectasis and parenchymal damage.[2] Over 85% of individuals have pancreatic insufficiency, which can result in significant malnutrition. Infertility is found in almost all men with CF. In addition, many individuals develop other complications including CF-related diabetes, chronic sinus disease, osteoporosis and cholestatic liver disease.[1]

Life-long, burdensome therapies are required to maintain health and extend survival.[3] Most individuals with CF require a high-calorie diet and pancreatic enzyme supplementation to combat malnutrition and maintain an adequate body mass index. Pulmonary therapies include

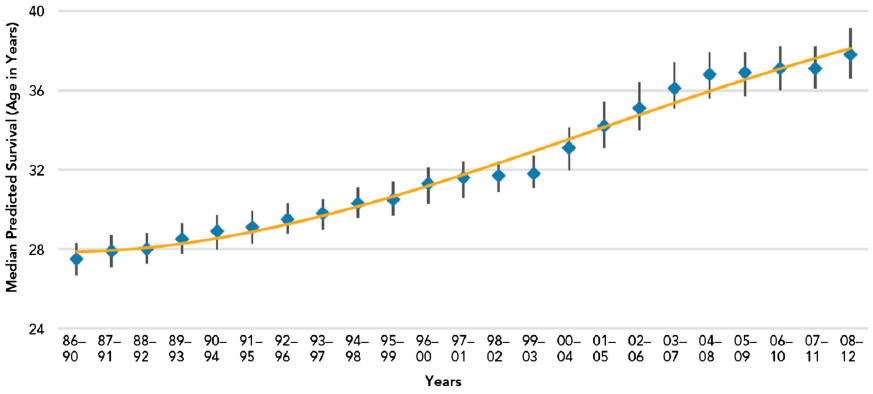

Figure 1 Median predicted survival for individuals with cystic fibrosis (CF) 1986–2012 in 5-year bands. The median predicted age of survival in 2012 is 41.1 years (95% CI 37.4 to 43.1). By using 5-year bands, the year-to-year variability decreases and the confidence bounds narrow. The median predicted survival for 2008–2012 is 37.8 years (95% CI 36.6 to 39.1). Data derived from the CF Foundation Patient Registry.[1]

daily airway clearance therapy and inhaled medications to thin secretions and control chronic infections. As lung disease progresses, daily symptoms of breathlessness, cough and sputum production increase, and the required therapeutic regimen becomes progressively more complex and time consuming.

Pulmonary exacerbations, which increase in frequency with disease progression, are characterised by an acute increase in respiratory symptoms and decrease in lung function. Pulmonary exacerbations are treated with oral, nebulised and/or intravenous antibiotics, more frequent inhaled and airway clearance therapies, and increased calorie intake.

Preventative therapy and early identification and treatment of symptoms, including worsening nutrition status (eg, decreasing body mass index), signs of worsening pulmonary disease and complications are key to maintaining health and quality of life. The complex and time-consuming nature of the therapies needed to optimise lung function and nutrition, as well as their high cost, place a tremendous burden on individuals with CF and their families. In addition, depression and anxiety are common issues for both individuals with CF and their families. For these reasons, adherence to prescribed therapies can be challenging.

CF CARE CENTER NETWORK

The CF Foundation has been at the forefront of identifying and fostering the elements that would allow individuals with CF to obtain optimal care. It was recognised early that experts from a variety of disciplines were required to effectively care for people affected by this multisystem disease. Therefore, in 1960, the CF Foundation established the Center Committee with the mandate to establish criteria for care centres that provide specialised care for individuals with CF and support for their families. Along with providing expert clinical care, the Center Committee stipulated that a comprehensive CF centre should also develop basic and clinical CF research expertise and educate the next generation of providers.

The Center Committee developed standards for care, research and teaching, which became the basic criteria that care centres must implement and sustain for CF Foundation accreditation. The initial standards of care required that CF care centres use a team of multidisciplinary experts to address inpatient and outpatient clinical needs, including psychological and sociological impacts, adhere to care practices developed by CF clinical experts, and maintain a sufficient

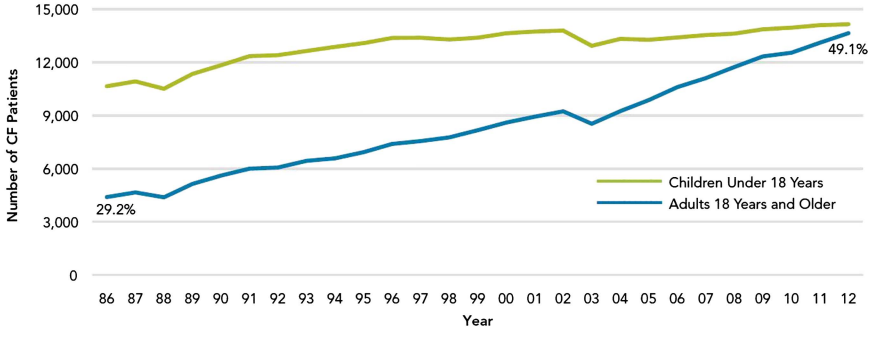

Figure 2 Proportion of individuals with cystic fibrosis (CF) who are adults. The change in the adult population from 1986 to 2012. Data derived from the CF Foundation Patient Registry.[1]

number of individuals with CF to advance the expertise of the centre's care providers.

At the recommendation of the Center Committee, the CF Foundation accredited its first two care centres in 1961 based on the newly established criteria. Accreditation of 30 more centres followed over the next 4 years, with the majority of care provided in paediatric hospital settings. By 1980, most of the current 115 centres in the Care Center Network were accredited. To ensure that care centres sustain accreditation standards, they undergo a site visit by members of the Center Committee at least every 5 years.

To share the growing body of knowledge and promote provider education, the CF Foundation sponsored the first North American CF Conference (NACFC) in 1986. This annual conference is the major scientific and clinical CF conference in the world, which has offerings for all disciplines involved in CF care. Each discipline is represented on the Program Planning Committee, and discipline-specific sessions are held throughout the conference. The NACFC is important in building and sustaining relationships across the Care Center Network, and disseminating quality improvement (QI) strategies, tools and successes.

Survival for people with CF has steadily improved, resulting in a growing population of adults with the disease. Now, nearly half of all CF patients are adults (figure 2).[1] This demographic shift necessitated the establishment of standards for the care of adults with CF. In 1997, accreditation criteria were amended and now require that care centres provide care for adults with CF in age-appropriate facilities, by physicians who are board-certified in adult care. By the end of the year 2000, to maintain CF Foundation accreditation, centres that cared for 40 or more adults were required to have separate paediatric and adult programmes staffed by care teams with age-appropriate training and expertise. With this change, paediatric and adult care programmes work collaboratively to develop and implement a formal paediatric-to-adult transition plan to coordinate care as well as teaching and research activities.

Not since the advent of the Care Center Network has the challenge to provide the necessary elements for CF care, research, education and thus accreditation been as great. Challenges have included the lack of internists or family practitioners experienced in adult CF care, the reluctance of paediatricians to relinquish the long-term relationship since the CF diagnosis, and resistance from many adult patients and families who did not wish to change providers and members of their care team.

In 2000, only a few of the larger, university-affiliated centres were providing care to adults within models that conformed to the new CF Foundation adult care criteria. Many care centres struggled with the proposed change—some in principle and some in securing the necessary resources to make the change. To facilitate the development of adult programmes, the CF Foundation created grants to help support training and recruitment of adult-trained providers to CF care. Today, while nearly all care centres include an accredited adult CF care programme, meeting all CF adult care standards continues to be a common challenge identified during accreditation site visits.

While the general accreditation standards are the same for all centres, specific criteria (eg, expectations for teaching healthcare professionals) are adapted depending on settings—university and community hospitals as well as private practices. This adaption helps to ensure that all individuals with CF are receiving appropriate care within a reasonable distance from home. By facilitating age-appropriate care within these settings, the CF Foundation care model provides a unique, sustainable approach to management of a complex chronic disease throughout the life span.

The goal of the accreditation process is to not only ensure that a minimum standard is being adhered to, but also promote optimal care. For this reason, the accreditation process supports and fosters continuous improvement efforts within care centres. The expectation that each care centre have a QI programme in place was added to the accreditation and oversight process in 2004.

The mission of the Center Committee is to foster exemplary care for all individuals with CF through the promotion of standards of care, accreditation of care centres, education of providers, and the advancement of research in all aspects of CF. The Center Committee comprises 20 paediatric and adult care physicians from across the USA who serve as CF Program Directors within the Care Center Network. It also serves as a valued advisory committee to the CF Foundation, contributing to the relevancy of the work of the CF Foundation and its accredited care centres.

IDENTIFYING THE IMPROVEMENT OPPORTUNITIES

The creation of the CF Foundation Patient Registry in 1966 was a key advancement in the ability to achieve improvements in CF health outcomes nationally. This data tool tracks the impact of CF care standards on the health of those living with the disease. The Patient Registry became a web-enabled database, known as Port CF, and is deployed throughout the entire Care Center Network as an institutional review board-approved, longitudinal, observational study. Providers can enter the health data for all CF patients at the care centre who have consented to participate. The Patient Registry provides key longitudinal epidemiological data and reports for care centres and the CF community to enhance the understanding of the natural history of the disease and help identify complications and emerging respiratory pathogens.[1] Multivariate modelling of

data from the CF Foundation Patient Registry has identified several factors that influence survival and other important clinical outcomes. Factors known to affect survival, such as lung function, nutritional status, airway microbiology, CF-related diabetes and adherence to chronic therapies, are important targets for QI efforts (table 1).

Fifty years of steadily improving survival is a source of pride for CF Foundation senior leadership. It demonstrates their commitment to achieving the mission of improving the quality of life for everyone with CF. In 1999, the data from the Patient Registry were first analysed at the centre level. As described by Schechter *et al*,[24] data analysis exposed marked variation in key outcomes, including survival, across the Care Center Network. For example, examination of data from 1990 to 2001 at the seven care centres with the best CF health outcomes demonstrated a remarkable survival gap between these care centres and the rest of the Care Center Network (figure 3). People cared for at those seven centres were expected to survive 6.5 years longer than individuals at the rest of the network. CF Foundation leadership and care centre providers began asking why some centres had better survival when all used the same standards for CF care.

HAVING THE WILL TO CHANGE

The CF Foundation leadership felt an urgency to address the variation in outcomes and close the gap between the care centres with the best outcomes and the rest of the centres. The goal was to improve care for everyone with CF in the USA by embracing and spreading QI processes and principles for the delivery of care within the accredited Care Center Network. It was clear to the CF Foundation leadership that additional years or even decades could be added to the lives of those with CF by enhancing care delivery.

The CF Foundation's leadership started by reaching out to national healthcare quality experts and leading

Table 1 Selected factors that influence lung function or survival in individuals with cystic fibrosis (CF)

Parameter	Reference
CF-related diabetes	4, 5
Exacerbations	6
Gender	7–9
Insurance status	10
Infection with *Pseudomonas aeruginosa*	7, 11–13
Infection with methicillin-resistant *Staphylococcus aureus*	14, 15
Infection with *Burkholderia cepacia complex*	16, 17
Malnutrition	7, 18, 19
Pancreatic insufficiency	7, 8, 13
Poor adherence	20
Second hand cigarette smoke exposure	21
Socioeconomic status	10, 22

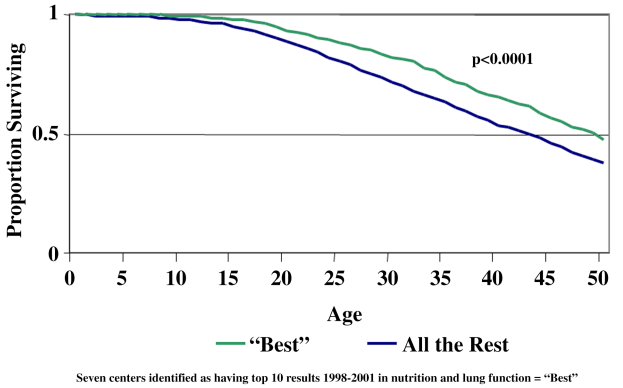

Figure 3 Survival at the seven care centres with the best cystic fibrosis (CF) health outcomes (green line) compared with the rest of the Care Center Network (blue line) from 1998–2001.

physicians within the network and developed a strategic plan for improvement. While not unique to CF, the key strategies and opportunities for improvement were presented to and agreed upon by the CF care centre leaders in 2003 (box 1). The QI strategies include developing leadership—nationally and within each care centre—identifying best CF care practices, and incorporating people with CF and their families into the improvement effort. Every care centre, no matter its size, could adopt these strategies and participate in improving the quality of care for people with CF at their care centre and thus across the USA. With the Patient Registry and the growing body of data, it was now possible to track outcomes and practices at individual care centres.

Initially, some care centre directors, physicians and team members were hesitant to believe that their care centre did not have the best possible health outcomes. People believed they were doing their best. Some had to overcome their inclination to doubt the validity of their centre's Patient Registry data. They had to get past the belief that the care they delivered was the 'best' and that the data were wrong. The CF Foundation worked to help care teams understand their data and develop approaches to use it and improve outcomes. The data were for learning and improvement. Providers were assured that the data would not be used punitively for suboptimal

Box 1 Strategies and opportunities for improvement in cystic fibrosis (CF) care

1. Developing and sustaining leadership for change
2. Sharing quality improvement tools and approaches with all CF care centres
3. Incorporating people with CF and their families into the improvement work
4. Identifying and enabling CF care 'best practices'
5. Providing decision support for CF care teams

outcomes, but as a metric to continue improvement in the health outcomes of people with CF. In contrast, some care centres were early adopters and quickly understood the importance of, and how to use, the Patient Registry data to identify areas where they could improve clinical practice and processes and thus the quality of CF care they delivered.

Transparency of the care centre level data from the Patient Registry became publically available for the first time on the CF Foundation's website (http://www.cff.org) in 2006, followed by a 2-year planning process. The public data is a key driver for improvement for the care centre staff, people with CF and their families and institutions. In addition, data transparency opened the door for further incorporation of people with CF and their families into the improvement work and as full partners in CF care.

CHANGING THE CULTURE

The vision of the CF Foundation leadership is that exemplary care should be delivered at all CF Foundation-accredited care centres to further extend the quality and length of life for people affected by the disease. Simply developing standards of CF clinical care and disseminating the health outcome data is not sufficient to improve care—practical strategies are required to achieve improvement. Along with the development of key strategies and opportunities for improvement (box 1), seven 'worthy goals' for CF were developed to provide a framework for care centres to strive toward (box 2). Over time, multiple improvement strategies have been developed and implemented by the care centres and continue to be developed and shared generously throughout the network.

The CF QI work was launched with the development of yearlong 'learning and leadership collaboratives' (LLCs) designed to teach QI principles to CF care teams and provide an environment for learning from peers at other care centres and QI coaches. Other initiatives included benchmarking of paediatric and adult CF programmes with the top health outcomes. Through these initiatives, key drivers influencing high-quality care were identified, compiled and shared throughout the Care Center Network.

Revising CF care standards to guidelines based on evidence published in the medical literature was another identified strategy for improving CF care and health outcomes. These evidence-based care guidelines, which are published in peer-reviewed journals, provide caregivers with standards with which they can compare and improve their practice patterns and processes.[23] Discipline-specific mentoring programmes that partner experienced CF multidisciplinary team members with those new to CF care—sharing knowledge, tools and experience across the Care Center Network—are very successful. New physicians have been recruited to CF care through award programmes

Box 2 Seven worthy goals

1. Patients and families are full partners with the cystic fibrosis (CF) care team in managing this chronic disease. Information and communication will be given in an open and trusting environment so that every patient/family will be able to be involved in care at the level they desire. Care will be respectful of individual patient preferences, needs and values.
2. Children and adolescents will have normal growth and nutrition. Adults' nutrition will be maintained as near normal as possible.
3. All patients will receive appropriate therapies for maintaining lung function and reducing acute episodes of infection. Pulmonary exacerbations will be detected early and treated aggressively to return patients to previous levels of lung function.
4. Clinicians and patients will be well-informed partners in reducing acquisition of respiratory pathogens, particularly *Pseudomonas aeruginosa* and *Burkholderia cepacia* complex.
5. Patients will be screened and managed aggressively for complications of CF, particularly CF-related diabetes.
6. Severely affected patients will be well supported by their CF team in facing decisions about transplantation and end-of-life care.
7. Patients will have access to appropriate therapies, treatments and supports regardless of race, age, education or ability to pay.

focused on increasing opportunities related to both research and clinical practice. Initiatives to build and strengthen the partnership between people with CF, their families and care centre staff, such as patient and family advisory councils, have also been used to drive improvement. All care teams participating in an LLC are expected to include a person with CF and/or family member in the lead improvement team. In 2006, only 64.5% of care centres involved people with CF or families in the centre's QI work through surveys, initiation of advisory boards, etc. Today over 85% of care centres involve their patients and families in improving CF care locally. The most dramatic change was observed in the development of patient or family advisory boards. The number of care centres with an advisory board has increased from 31 in 2006 to 97 in 2012.

To continue the vision of exemplarily care and outcomes, all care centres are now mandated to incorporate continuous QI efforts into their clinical practice. The value of these programmes to people with CF and the CF Foundation is demonstrated by the fact that QI work in the CF care centre and the positive health outcomes reported in the Patient Registry are now part of the care centre accreditation evaluation.

CONCLUSION

The CF Care Center Network is a model of chronic care and continuous improvement in health outcomes. The CF Foundation has developed an integrated approach to providing multidisciplinary care that includes the Care Center Network, Patient Registry and clinical practice guidelines, which have provided fertile ground for QI initiatives. The success of the Care Center Network is reflected in the improvement of lung function, nutritional status and survival for individuals with CF. The CF Foundation's QI programme demonstrates that providing leadership and a framework for dissemination of data, knowledge, tools and expertise can improve care for a complex chronic disease at multiple clinical care sites across the USA. However, this approach is only possible with a strong partnership between patients, families and the multidisciplinary healthcare providers needed to care for individuals with CF.

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Notes