Journal of Cystic Fibrosis 23 (2024) 1055–1065

Contents lists available at ScienceDirect

Journal of Cystic Fibrosis

journal homepage: www.elsevier.com/locate/jcf

Original Article

Cystic fibrosis foundation position paper: Redefining the CF care model

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ARTICLE INFO

Keywords:
Cystic fibrosis
CF care model
Healthcare delivery
Telemedicine
Mental health

ABSTRACT

Specialized care is provided to people with cystic fibrosis (pwCF) by interdisciplinary teams nested within the CF Foundation's accredited care center network. This network allows for standardization of the care model, implementation of clinical care guidelines, efficient communication, and outcomes reporting. Recent developments have impacted this care model. Increased access to CFTR modulator therapies has improved overall health for many, although not all pwCF. The COVID-19 pandemic resulted in a rapid adoption of telemedicine and remote monitoring to ensure continuity of CF care. A collaboration of care providers, pwCF, and parent caregivers reevaluated key aspects of the current care model and considered potential modifications based on a widening range of needs. Available evidence was used to evaluate components of routine clinical practice and identify potential adaptations to care. The review included identification of patient characteristics warranting intensive monitoring, while embracing patient-centric care, and emphasizing the integration of telemedicine and at-home health technologies. Despite the changing landscape, the importance of the relationship between pwCF, their support system, and the care team was confirmed as a timeless and foundational aspect of the care model. Shared decision making, partnership, and coproduced care plans between pwCF and their CF care teams guide the best adaptations of the care model to support individual priorities and wellbeing. As health care advances and pwCF age, further research is needed to understand the impact of the care model on long-term health outcomes and to identify best practices that support pwCF to live longer healthier lives.

Abbreviations

AAP	American Academy of Pediatrics
ACFLD	Advanced CF lung disease
aCFLD	Advanced CF liver disease
BAL	Bronchoalveolar lavage
CXR	Chest x-ray
CF	Cystic fibrosis
CFF	Cystic Fibrosis Foundation
CFRD	Cystic fibrosis related diabetes
CFTR	Cystic fibrosis transmembrane conductance regulator
CT Chest	Computed tomography of the chest
DXA	Dual energy x-ray absorptiometry
EMR	Electronic medical record

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https://doi.org/10.1016/j.jcf.2024.08.007
Received 25 June 2024; Received in revised form 14 August 2024; Accepted 30 August 2024
Available online 25 September 2024

ETI	Elexacaftor-tezacaftor-ivacaftor
GAD-7	Generalized Anxiety Disorder-7
GI	gastrointestinal
HRCT chest	High resolution computed tomography of the chest
HRSN	Health-related social needs
IVA	Ivacaftor
LCI	Lung clearance index
MBW	Multiple breath washout
MRI	Magnetic resonance imaging
NTM	Non-tuberculous mycobacteria
PHQ-9	Patient Health Questionnaire-9
PA	*Pseudomonas aeruginosa*
PCP	Primary care provider
pwCF	People with CF
QI	Quality improvement
SDOH	Social determinants of health

1. Introduction

The hallmark of cystic fibrosis (CF) care is the interdisciplinary team approach in providing both surveillance for the development of disease progression and acute care of complications. Pioneered in the 1960s, the current CF care model was defined initially by clinical practice guidelines in 1997 [1]. The care model has evolved in response to advances in therapeutics, evidence-based recommendations [2], quality improvement (QI) initiatives, and epidemiologic data collected in a patient registry [3]. Guidelines specific to numerous aspects of CF care across the lifespan provide recommendations for visit frequency, screening tests, and treatment of CF-related health complications. This model of care has facilitated remarkable improvements in clinical outcomes such that the median predicted age of survival is 61 years for pwCF born between 2019 and 2023 in the United States [4].

For decades, CF therapies targeted health consequences of CF, but greater understanding of CF genetics led to the development and approval of CF transmembrane conductance regulator (CFTR) modulators that target the underlying cause of CF [5–7]. While CFTR modulators are not a cure, they have been transformational for many people with CF (pwCF), resulting in improved health. However, for pwCF not eligible, unresponsive to or intolerant of CFTR modulators, there remains a greater risk of disease progression and complications compared to pwCF on modulators. Treatment complexity and burden [8], along with unmet needs [9] unique to each person, may impact an individual's ability to manage their disease effectively. Commensurate with health improvements related to treatment advances, CF care teams have increased their focus on relationship building, shared decision making, coproduction of care, and supporting disease self-management through more systematic approaches to transition education [10] and skill development. There is also importance in identifying and addressing social drivers or inequities that affect health (i.e., ethnic, racial or cultural factors) [11–14].

Access to quality care is one of many factors that has a marked impact on health outcomes [15]. The CF care model, like the entire healthcare system, endured stress during the COVID-19 pandemic. One positive by-product of this challenge was increased availability of telemedicine and remote clinical monitoring as viable ways to support CF care [16–19]. Given the improvements in health resulting from CFTR modulators and new opportunities to harness technology in CF care, it is necessary to re-examine the CF care model, evaluating what is ideal monitoring to maintain optimal health and prevent complications for all pwCF. Goals include reducing potentially unnecessary healthcare utilization for those with reduced care needs, while maintaining a care model that continues to support those with advanced or complicated disease, and those who do not have access to or cannot tolerate CFTR modulators.

The CF Foundation (CFF) intends for this position paper to provide reasonable clinical guidance, based on available empirical evidence and expert opinion, to clinicians, pwCF, and other stakeholders. Care decisions should contextualize the guidance with an individualized risk-benefit assessment and the needs and priorities of the pwCF and family.

2. Methods

Through a request for applications, the CFF convened a geographically diverse interdisciplinary committee to critically appraise the current care model. Literature was reviewed to assess how CF care is delivered and identify opportunities for innovation and improvement. Clinical suggestions for evolving the care model, encompassing considerations for visit frequency, care settings, and health maintenance and screening were crafted collaboratively. This position paper integrates available evidence with expert experience to guide the evolution of CF care considering recent scientific and technological progress. This is not intended to reassess published guidelines regarding specific CF complications nor to address use of available medications. Rather, its purpose is to offer guidance for monitoring and surveillance in the era of advancements to therapies and technology. The scope of this position paper is limited to routine outpatient care. The unique needs of those patients with advanced lung disease or post-transplant are addressed in guidelines and consensus statements [20,21].

3. The CF care model

The original CF care guidelines established in 1997 [1] were primarily informed by expert opinions concerning visit frequency and health maintenance for pwCF. Because these guidelines have been associated with improved health outcomes over time, a conservative approach to changing the model was taken. When evidence strongly supported adjustments to the model, then modifications were considered to align with current needs and technological advancements. The proposed guidance supports a continued rigorous approach of periodic assessments to identify and manage early signs of clinical deterioration, but has identified characteristics of health stability in which less intense monitoring might be acceptable. In contrast, specific patient groups were identified that may be at higher risk of rapid health decline, and for whom reducing the frequency of clinical visits and health surveillance would not be appropriate (Table 1). Note that this guidance is independent of CFTR modulator use as great heterogeneity exists among pwCF regardless of CFTR modulators. Instead, these detailed delineations help tailor care more closely to individual patient risks and needs, reinforcing the model's foundation on patient-centric principles and evidence-based practice.

3.1. Visit frequency

Suggested visit frequency is based on age and health stability irrespective of modulator use (Tables 1 and 2).

3.2. Age-based considerations

Infants, toddlers and preschoolers should have frequent visits for education and relationship building, developing health literacy and skills for symptom monitoring, following current guidelines [22,23]. Surveillance for early infection with *Pseudomonas aeruginosa* (PA) is essential and may increase the success of eradication[40,41]. Another key driver for close monitoring of the young population is to be able to respond to rapidly changing needs related to growth, nutrition, and development. This will help optimize long-term pulmonary outcomes and overall health trajectory.

For children aged ≥6 years and adults, a decrease in surveillance visit frequency to every 4–6 months may be appropriate when health status in all domains is deemed stable (Table 3), and spirometry technique has been mastered. However, an in-person clinic visit no less than every 6 months should occur for monitoring, regardless of health

Table 1
CF populations who benefit from frequent CF care visits (at least every 3 months).

Population	Reasons for frequent care visits
New CF diagnosis (or new CF complication, e.g. CFRD or ABPA)	• Determine clinical phenotype and trajectory of disease • Identify acute stressors and offer tailored support for adjustment • Build relationships with clinical team • Focus on comprehensive education and development of self-efficacy
Infants and Toddlers (0–24 months)	• Provide anticipatory guidance • Determine clinical phenotype and trajectory of disease • Build relationships with clinical team • Focus on comprehensive education and development of self-efficacy • Establish routines and cooperation during key developmental window
Preschoolers (2–5 years)	• Provide anticipatory guidance • Initiation of CF treatments and assessment of response, along with laboratory studies • Focus on comprehensive education and development of self-efficacy • Key developmental window to establish routines, and psychosocial needs • Opportunity to strengthen relationships with the CF care team
Transition periods (i.e. adolescence and young adulthood, pregnancy, new parenthood, transplant, geographical move to new center, ongoing education needs, new employment, occupational demands, loss of support)	• Provide anticipatory guidance • Identify acute stressors (e.g. loss of employment) and offer tailored support • Determine effects of transition on health parameters, adjust medical management and assess response to intervention • Opportunity to strengthen relationships with the CF care team
Underrepresented pwCF that may benefit from more frequent in-person interactions	• Utilize interpreter for education and challenging conversations • Engage in culturally sensitive interactions • Provide written educational materials and/or demonstrations in person • Opportunity to strengthen relationships with the CF care team • Increase health literacy • Opportunity to access additional resources
Change in health status or at-risk health status (i.e., declining lung function, frequent pulmonary exacerbations, advanced lung disease, change in microbiology, disease progression, smoking/vaping)	• Implement new therapies or smoking cessation efforts and assess response to intervention • Discuss and refer for lung transplantation based on care guidelines • Opportunity to strengthen relationships with the CF care team
Unmet mental health or health-related social needs (HRSN) (i.e. unreliable access to medications or insurance)	• Identify needs and offer resources • Assess response to intervention • Monitoring via telephone or video-based telemedicine in between in-person visits may be beneficial

stability, in order to ensure confidence in clinical measures, promote wellness, medication safety and adherence. A CFF patient registry-based analysis demonstrated that individuals with gaps in CF care greater than six months had lower lung function compared to their age matched peers. Additionally, there was a direct correlation between longer gaps in care and greater lung function decline [42]. Although these data were collected prior to elexacaftor-tezacaftor-ivacaftor (ETI), it suggests that a reduction in clinic visit frequency should be approached cautiously. Given the dynamic and progressive nature of CF disease, early detection of declines in health or lung function would warrant the return to more

Table 2
Guidance for the CF Care Model.

Component of Care	Previous Guidance	Current Guidance
Routine clinic visit	Infants (0–6 months): at least every month [22] Infants (6–12 months): every 2 months [22] Toddlers (1–2 years): every 3 months [22] Preschoolers (2–5 years): every 3 months [23] Children ≥ 6 years and adults: every 3 months for everyone [24]	Infants (0–6 months): no change Infants (6–12 months): no change Toddlers (1–2 years): no change Preschoolers (2–5 years): no change Children ≥ 6 years and adults: every 4–6 months can be considered when health is determined to be stable in all domains (Table 3)
Interdisciplinary assessment (annual review of each pwCF by core care team members)	At least annually Adult: at least annually [24]	No change Genetic counselor, as needed (see Care Team Position Paper) Pharmacist, as needed (see Care Team Position Paper)
Immunizations: Work with PCP	Primary immunizations: given at PCP, as per American Academy of Pediatrics guidelines Influenza: annually [22–24]	Primary immunizations: no change Influenza: no change COVID-19: annually
Immunizations	RSV: RSV monoclonal antibody (palivizumab): Infants with CF <12 months [22]: monthly during RSV season	RSV: Infants/Toddlers: RSV monoclonal antibody (nirsevimab), preferred: once before 8 months of age during RSV season (Nov-Apr) RSV monoclonal antibody (palivizumab), unchanged: monthly during RSV season if nirsevimab not available Adults age ≥ 60: once during RSV season (prefusion F glycoproteins) Pregnant women 32–36 weeks gestation: once during RSV season and/or nirsevimab to infant as above
	Pneumococcal vaccine: as per CDC guidelines [25]	Pneumococcal vaccine: No change [25]
Routine pulmonary function testing	In-person at CF clinic: Children 3–5 years: attempt at least annually; not always feasible [23] Children ≥ 6 years: at least twice yearly, preferred every 3 months (quarterly) [26] Adults: regularly monitored and performed on most visits [24]	In-person at CF clinic: Children 3–5 years: No change Children ≥ 6 years and adults: at least twice per year (with every clinic visit)
	At-home or at local facility providing spirometry: • Began during pandemic and used variably among centers	At-home or at local facility providing spirometry: • Adjunctive, between clinic visits or with telemedicine visits as needed • May be useful if barriers to travel to CF center • May be challenging to document in EMR or to

(*continued on next page*)

Table 2 (*continued*)

Component of Care	Previous Guidance	Current Guidance
		use clinically in children
Exercise assessment	6-minute walk test for ACFLD and transplant referral[20,27]	No change
Imaging	CXR: Infants and Toddlers (0–24 months): within 3–6 months old and at age 2 [22] Children > 2 years of age: every 1–2 years [23] Adults: every 2–4 years or as needed [24]	CXR: No benefit to use of CXR for surveillance
	CT chest: consider in infants and preschoolers or at any age, or when needed to assess lung disease[22,23,28]	CT chest: *strongly* consider in infants and preschoolers or any age with clinical need to assess and preferentially in place of CXR, may repeat if clinically necessary at clinician's discretion[22,23,28]
	Abdominal ultrasound: not routine prior to 2024hepatobiliary guidelines [29]	Abdominal ultrasound: Age ≥ 3 years until late adolescence every 2 years and a baseline in adults: [29]
	Bone density: Age ≥ 18 years with CF: baseline DXA scan pwCF ≥ 8 years with risk factors for osteoporosis: repeat every 1–5 years based on result [30,31]	Bone density: no change
Respiratory specimen cultures	Sputum (preferred) or oropharyngeal (OP) swabs: [22-24] Infants (0–24 months): every 2–3 months Preschoolers (2–5 years): every 3 months Children ≥ 6 years to adults: every 3 months	Sputum (preferred) or oropharyngeal (OP) swabs: Infants and Toddlers: no change Preschoolers (2–5 years): no change Children ≥ 6 years to adults: no change, consider feasibility of home and local monitoring
Laboratory/other evaluations	Hematology, chemistry, IgE, liver function: All pwCF: annually[24,29]	No change
	Fasting or non-fasting serum lipid levels: Children: consider screening for familial dyslipidemia at least once during childhood and more regularly if cardiovascular risk factors Adults: annually as per primary care guidelines [32,33]	No change
	• Blood pressure: at least annually at clinic visits in children 3–17 years [34] and annually in adults ≥ 40 years or with increased risk; adults aged 18–39 years without risk factors can be screened every 3–5 years [35]	No change
	Fat-soluble vitamin levels: Infant: approximately 2 months after starting vitamin supplementation and annually thereafter [22]	Fat-soluble vitamin levels: Infant: approximately 2 months after starting vitamin supplementation and annually thereafter,

Table 2 (*continued*)

Component of Care	Previous Guidance	Current Guidance
	Preschoolers [2-5]/Children: Annually with more frequent measurements after dose adjustment [23]	plus recheck following initiation of CFTR modulators[36,37] or after adjustments to vitamin supplementation
	Adult: Serum levels or retinol, vitamin E, and 25-hydroxyvitamin D should be checked annually [24]	Preschoolers [2-5]/Children: Annually with repeat measurements after dose adjustment [23], plus recheck following initiation of CFTR modulators[36,37] Adult: Serum levels or retinol, vitamin E, and 25- hydroxyvitamin D should be checked annually [24], plus recheck following initiation of CFTR modulators[36,37] or after adjustments to vitamin supplementation
	Fecal elastase: at diagnosis and when clinically indicated [22]	Fecal elastase: at diagnosis and reassess as clinically indicated. Assess again after starting modulators (especially if modulators were started in childhood). Optimal frequency and timing is unknown
CFRD screening (OGTT)	Children ≥ 10 years to adults: annually, or with clinical signs and symptoms of CFRD [38]	No change
Mental health screening	Children ≥ 12 years to adults: annually and offered to caregivers per center processes [39]	No change, in-person screening preferred
Health-related social needs screening	Not mentioned in a CF Foundation guideline	As part of annual social work assessment

This guidance is for routine clinical monitoring only and excludes acute indications or follow up of an intervention when additional testing may be appropriate.
Abbreviations: ACFLD=advanced CF lung disease.

frequent visits as circumstances change.

3.3. Assessment of health stability

When considering reducing clinic appointment frequency to every 4–6 months for children 6 years and older and adults, stability should be holistically evaluated (Table 3). Because time between visits may be longer, reliable self-advocacy and the ability to communicate with the CF care team in a timely fashion must be in place.

Reduced frequency from quarterly visits is not recommended for specific populations in whom there is less confidence in health stability, unreliable access to medications or insurance, and cultural nuances or language barriers (Table 1). Individuals with a new diagnosis of CF after age 6 will benefit from a minimum of quarterly visits until clinical phenotype and disease trajectory are well established and a period of health stability is reached. This allows for identification of unique health needs, development of self-efficacy and self-care skills.

There are several other transition periods in the lives of pwCF that may pose health challenges [10,43,44] during which continued quarterly, or even more frequent visits, may be indicated, including, but not limited to, adolescence, pregnancy and new parenthood, and advanced CF lung disease. The adolescent population has historically

Table 3
Factors to consider in holistic assessment of stable health.

Health Domains	Factors
Lung Health	• No clinically significant respiratory symptoms • No significant decline in lung function • No recent exacerbations • No new respiratory pathogen(s) • No significant changes/escalation/de-escalation of chronic therapies (including initiation of modulators) • No significant change in physical stamina or exercise tolerance
GI Health	• No significant changes in symptoms • Minimal or no medication adjustments • No new GI diagnoses
Endocrine Health	• No new diagnosis of CF related diabetes • Well-controlled CF related diabetes
Nutritional Health	• No significant change in weight or BMI • Not significantly underweight or overweight • No significant changes in diet pattern, vitamins, body image disturbances, etc.
Mental Health	• No significant increase in symptoms • No high-risk behaviors • Adequately addressed social needs • Appropriate access to mental health services outside of the CF team as needed
Social Health	• No unaddressed social needs such as: under or uninsured, limited access to therapies, food insecurity, etc. • No significant issues accessing care or medications • No significant barriers to communicating with team (language or other)

Legend: Factors that may be considered in determining that a pwCF has achieved health stability. If any of these factors are clinically significant, then a discussion with pwCF and family is warranted to support greater visit frequency.

demonstrated the most volatile changes in lung function[42,45–47]. Adolescent health is affected by biopsychosocial factors including puberty, increased independence in self-care, peer influence, and risky behavior. Preparation for transition to independent care in young adults requires incremental education to help pwCF gain knowledge, independence, and confidence in the maintenance of their own present and future health [10], including reproductive health. Pregnancy and starting a family are other times in life that could benefit from frequent visits for monitoring, education and support [48]. Occupational demands, including business travel and inflexible work hours, have the potential to supersede health promoting activities in adults, which can jeopardize clinical stability.

People with ACFLD represent a vulnerable population for whom at least quarterly evaluation is appropriate given the potential and time-sensitive need for discussion, referral and evaluation for lung transplantation [20]. After lung transplantation, care is shared between the transplant team and the CF team, with at least annual CF care team visits [21]. Less frequent clinic visits may result in longer appointment duration to allow completion of necessary testing and interdisciplinary assessments. Lengthy clinic visits may challenge the ability of pwCF and their families to remain engaged and should be considered in the shared agenda setting between the pwCF and their CF team. While having the entire team present at an appointment may appear more efficient, it could dilute learning and distract from the highest priority aspects of the visit. Asynchronous care by interdisciplinary team members via telemedicine may help reduce this burden if team coordination and communication can be maintained [49]. Along with the benefits of telemedicine, there can be challenges including barriers to access, licensing, reimbursement and insurance coverage [50–52] (Table 4). PwCF and their families may be more accepting of telemedicine than the CF team, and support may be due to the heterogeneity among CF centers in the ability to address barriers. Additional touchpoints using telemedicine between in-person visits may allow CF centers to enhance care for pwCF especially if the in-person visit frequency is two or three times per year. All reductions in visit frequency, and/or shift to telemedicine

Table 4
Telemedicine benefits and barriers [19,50–54].

Benefits	Barriers
Improved access to care and/or more frequent contact	Disparities in broadband internet access, accessible devices, appropriate translation services, comfort/ease with technology
Reduced time away from school/work	Institutional, state, and federal barriers to telemedicine delivery (legal and regulatory obstacles)
Reduced travel costs/increased convenience for those who live far from clinic	Possibilities for technical difficulties
Decreased infection risk	Limitations in performing comprehensive physical examination
Improve patient engagement	Limitations in ability to perform testing (spirometry, respiratory culture, imaging, blood draws) at the time of the visit
Opportunity to see the home environment and more comfortable at home	Privacy and security risks
Possible improved adherence to visit frequency	Reimbursement issues
Improve efficiency of visit	Decreased opportunity for teaching students/residents; increased follow up work outside of visits

should be a result of thoughtful, personalized, shared decision-making between the pwCF/family and their CF care team.

3.4. Maintenance health screening

3.4.1. Pulmonary function testing

Pulmonary function testing, specifically spirometry, is a critical outcome measure for monitoring lung health in pwCF. Even when visit frequency is decreased, spirometry should be performed in clinic at minimum twice yearly in pwCF ≥6 years old (Table 1) to establish trends. There are circumstances in which more frequent assessment of lung function may be useful to guide clinical decision-making. These include, but are not limited to, the addition or withdrawal of therapies, increased symptoms, declining lung function, documentation of a return to baseline lung function following exacerbation treatment, exercise intolerance or reduced stamina, or when a new, significant respiratory pathogen has been identified. Proficiency in spirometry technique is essential for accurate measures of lung function. Prior to considering home spirometry, it is essential to ensure pwCF can successfully perform this test to meet quality standards [55]. During this technique learning period, and, and throughout performance of home spirometry, it is essential to have consistent and trained staff to coach and reinforce proper technique.

Collaboration with local pulmonary function labs and home spirometry should be considered as adjunctive testing, particularly for those who live far from clinic, cannot easily access in-person testing, or when home spirometry is not an option. With proper patient training, home spirometry can be performed with accuracy and reproducibility, even in children [56], though values are notoriously lower than those obtained in the clinic[57,58]. Ongoing studies on the use of home spirometry include OUTREACH, an observational study comparing home to office spirometry in children 6 years of age and older and adults with CF [59]. Quality improvement (QI) studies are ongoing, focusing on logistics of home spirometry including accurate height measurements, technique and coaching for optimal results, and incorporation of results into overall care and the electronic medical record (EMR). With validation over time, home spirometry holds promise as an important remote monitoring tool in times of sickness and in health [24].

Spirometry is not the only measure of lung health and may not detect changes of early lung disease [60]. Lung Clearance Index (LCI), or multiple-breath washout (MBW), and impulse oscillometry offer additional assessments of lung function but are not routinely available at all

centers. Both are areas of ongoing research.

Exercise assessments to evaluate overall lung and cardiovascular health are used more commonly in patients with ACFLD [61] as reduced aerobic capacity may be associated with mortality in CF [62]. The role for formal exercise testing in the ACFLD population is addressed in other guidance[20,27]. Discussion of type and duration of exercise in everyday life will likely be of increasing importance given the holistic health benefits, particularly with aging [63].

3.4.2. Imaging

The low sensitivity and specificity of chest x-ray (CXR) limit its influence on clinical management [64] aside from acute changes (e.g. pneumothorax, or new opacity) and is not recommended for routine monitoring. Computed tomography of the chest (CT chest) is a more sensitive imaging modality for the detection of early bronchiectasis and mucus plugging. The merits of performing surveillance CT chest in the very young remain to be demonstrated. Prior to modulators, there was data that early lung disease or bronchiectasis could be identified in the asymptomatic patient [65]. The impact of early modulator use on this finding is an area of research interest. Another area of interest is to determine the appropriate cadence for subsequent imaging and its impact on treatment decisions [66]. As pwCF age, there is an increased prevalence of non-tuberculous mycobacteria (NTM) disease and now that many on modulators are not producing sputum for surveillance testing, it will be important to learn whether and when intermittent imaging is appropriate.

Magnetic resonance imaging (MRI) chest is a sensitive, non-ionizing radiation measure that can detect early structural CF lung changes and measure ventilation/perfusion (hyperpolarized xenon) [67–69]. MRI will continue to be researched in CF due to lack of radiation, advancing technology and increasing lifespan [70].

CF related liver disease is a leading cause of non-pulmonary mortality and can result in increased hospitalizations, declining nutritional health, and endocrine and bone disease [71]. Per the 2024 CFF guideline, abdominal ultrasound is recommended every two years for pwCF age $\geq$3 years until late adolescence and as a baseline test in adults to evaluate for CF hepatobiliary disease [29] (Table 1). Those with advanced liver disease undergo gastroenterology (GI) evaluations with liver elastography and cancer screening yearly as per the guideline [29].

3.4.3. Microbiology

Persistent airway infection is a common and serious feature of CF lung disease associated with accelerated lung function decline and bronchiectasis [72]. CF respiratory cultures are currently recommended quarterly in pwCF [22–24,73,74] (Table 2), but this could be challenging in the context of decreased visit frequency for those who meet criteria (see above). Less frequent CF respiratory cultures may compromise sensitivity in the detection of clinically significant pathogens; early identification and treatment with intent to eradicate PA has been proven to preserve lung function, improve symptoms, and lead to better quality of life [75]. Less frequent surveillance may lead to later detection and reduce the success of PA eradication [40,76]. Reducing the clinic visit frequency to 2–3x per year in healthy pwCF $\geq$ 6 years may pose challenges to obtaining quarterly microbiological specimens. Submitting specimens to a local facility may be acceptable if the lab adheres to CFF microbiology guidelines [73]. The utilization of mail-in specimens obtained at home and sent to the CF Center hospital laboratory has also been investigated with reliable results [77]. QI efforts are ongoing to assess the results of respiratory cultures obtained prior to or apart from clinic visits, and how this might impact clinical decision-making. However for many centers, cultures obtained in CF clinic and submitted to the CF microbiology laboratory may be the only acceptable method for surveillance cultures at this time.

Treatment with CFTR modulators decreases sputum production for many pwCF [78] resulting in fewer sputum cultures as well as less frequent detection of CF pathogens [79,80], such as PA [81]. However, reduced prevalence of pathogens on cultures or burden of infection does not equate to complete eradication of pathogens from the airways [82–84], nor is there evidence that eradication protocols or suppressive antibiotics are no longer beneficial in people on CFTR modulator treatment. Detection of respiratory pathogens is influenced by the sample source. OP swab cultures do not correlate well with sputum or bronchoalveolar lavage (BAL) culture results [85]. However, because OP swabs have a high negative predictive value for PA, more frequent OP swab cultures are beneficial to "rule out" the presence of PA [86]. Even with their limitations, OP cultures should be obtained in adults and children who cannot expectorate mucus. Induced sputum is another effective way to obtain cultures [87], though further research on possible implementation in clinic and home settings will be necessary. The absence of reliable sampling of sputum may influence the use of imaging to help determine when alternative methods of obtaining respiratory specimens (e.g. bronchoscopy) might be warranted.

An existing guideline recommends at least annual surveillance for NTM in those who can spontaneously expectorate sputum as well as for those who have clinical features of NTM infection [88]. The reduction in sputum production presents challenges for NTM surveillance as OP swabs are not acceptable for the performance of NTM cultures [88]. Alternative detection methods for NTM Screening in CF (induced sputum or bronchoalveolar lavage culture vs. other novel non-culture-based techniques) is an area for additional research. As noted above, imaging may be helpful in those who are unable to produce sputum.

3.4.4. Nutritional assessment

Optimal nutritional management remains an essential aspect of CF care, as nutritional status correlates with pulmonary outcomes and survival [89]. The impact of GI symptoms on pwCF can be profound, even in those on modulators. CFTR modulator treatment has resulted in less malnutrition but also a rise in rates of pwCF who are overweight or obese [90,91]. A recent CFF position paper on nutritional management outlines revisions to the traditional CF high fat/high calorie diet [92]. Although treatment with CFTR modulators appears to improve absorption of nutrients in pwCF, the need for pancreatic enzyme replacement therapy is largely unchanged to date in the CFF Patient Registry (CFFPR) [93]. As the age of CFTR modulator therapy initiation lowers, reassessing pancreatic exocrine function may be beneficial [92,94]. In modulator trials and case reports, some children with CF have recovered pancreatic function [95], and, while not as frequently, this has been seen in adults [94]. Therefore, assessment of fecal elastase is recommended as clinically indicated for all pwCF [96], at diagnosis and after starting CFTR modulators, especially in children. The long-term impact of CFTR modulators on pancreatic function is unknown, and the optimal frequency and timing for assessment of fecal elastase in pwCF on CFTR modulators is an area for further investigation.

The components of nutritional assessment vary based on age. For pediatric age groups, the focus is on dietary habits, healthy eating, growth and development, and adequate weight gain/maintenance. For older children and adults, the focus is on dietary habits, healthy eating, weight management, diabetes, and more recently, cardiovascular risk factors [97]. The need for accurate anthropometric measurements may require in person clinic visits for children, whereas home weights may provide sufficient information for adults. Determining alternate methods of assessing body composition is an area of ongoing research and likely will require in-person evaluation [98–100]. An assessment of a pwCF's body image and symptoms of disordered eating are important components of the dietitian's evaluation [92]. Salt replacement and hydration are relevant topics of discussion with pwCF, and the salt replacement need may not be as great in pwCF on modulators [92]. Nutritional management involves tailored dietary advice and individualized discussions with pwCF and their families to help them reach their personal goals, and to prepare young people for a healthy and extended adulthood [97].

Annual monitoring of fat-soluble vitamin levels (more often as clinically indicated) [92] and reassessment following initiation of CFTR modulators [36,37,101–107] are suggested with adjustment in fat-soluble vitamin replacement based on levels. The prevalence of obesity is increasing in pwCF, potentially changing the risk of cardiovascular disease[108,109]. Cardiovascular health assessment should be done in partnership between the CF care team and an individual's primary care physician (PCP), beginning at 3 years of age with annual monitoring of blood pressure (with age-appropriate technique, cuff and reference values) [110]. Consideration of screening for family dyslipidemia at least once during childhood and more regularly in children and adults with risk factors is suggested in accordance with general population guidelines [32,111,112]. A relevant research topic will be the impact of CFTR modulators on lipid levels and cardiovascular disease as pwCF age.

3.5. Cystic fibrosis related diabetes (CFRD)

CFRD is a common complication for pwCF, and its prevalence increases with age [113]. The CFRD guideline recommends annual screening with an oral glucose tolerance test in pwCF above age 10 years or with clinical symptoms prior, as hemoglobin A1c is not sufficiently sensitive for screening in pwCF [38]. Preliminary evidence suggests that CFTR modulators may preserve pancreatic endocrine function in some patients [114], which may delay or prevent CFRD [115]. However, CFTR modulators are associated with weight gain which may raise the risk of insulin resistance [115–118]. As such, CFRD screening should continue as per previous guidelines (Table 2). There may be opportunities to utilize other modalities such as continuous glucose monitor devices for CFRD screening as more data become available.

3.5.1. Bone health

Baseline bone density screening with dual energy x-ray absorptiometry (DXA) scan is recommended for pwCF over 18 years of age, and for pwCF over 8 years of age with specific risk factors for bone disease [30,31]. Testing should be repeated every 1–5 years, based on risk factors. The importance of strengthening and weight-bearing exercise in improving bone density is well known [119] and should be incorporated into care. Those pwCF with osteopenia or osteoporosis will need care from trusted partners including PCPs or endocrinologists. There is evidence of improvement in bone density in pwCF on ETI[120,121]. Further research on CF bone health is needed.

3.5.2. Cancer screening

CF confers a greater risk of colorectal cancer than the general population. Care recommendations include earlier screening initiation, more frequent screening frequency, and exclusive use of colonoscopy as the screening tool [122]. Research into a potential role for DNA-based stool testing for screening is ongoing, but not yet endorsed in CF given the high incidence of colon polyps, which require colonoscopy for detection and removal [123]. Although there is an increased risk of other GI cancers such as pancreatic and liver cancers among pwCF [124,125], there are no general CF specific screening guidelines. For those with advanced CF liver disease (aCFLD), children are screened yearly for hepatocellular carcinoma (HCC) with elastography and alpha-fetoprotein; adults are screened as per American or European Association for Study of Liver Disease guidelines [29].

Endoscopy may be warranted in pwCF due to higher rates of Barrett's esophagus and esophageal cancer, but exact data or guidance is lacking [126]. Cancer screening should be a part of transplant clinic follow up due to the additive increased risk for pwCF on immunosuppressive drugs [21,127]. Cancer screening in all pwCF, especially in those on immunosuppression for lung or other organ transplants, should adhere to published guidelines and should involve discussions with the PCP or specialist (OB-Gyn, Urology, Transplant Team, Dermatology) [24]. There is no evidence that CFTR modulator therapy reduces the risk of cancer. In fact, with increased life expectancy, cancer screening becomes of greater importance.

3.5.3. Mental health screening

The CF depression and anxiety screening and treatment guidelines recommend annual screening for pwCF aged 12 years through adulthood (Table 1) with the Patient Health Questionnaire-9 (PHQ-9) and the Generalized Anxiety Disorder-7 (GAD-7) [39]. The guideline includes recommendations for follow-up screening and targeted intervention until symptom scores are in the minimal range. There is evidence that intervention can improve symptoms and buffer against the negative impact of disease or non-CF related acute stressors [128,129].

Comprehensive mental health screening also includes consideration of other patient-reported outcomes including quality of life, common mental and behavioral health comorbidities such as pain, substance use, sleep, executive functioning challenges, body image concerns, and disordered eating [128]. Screening can ensure pwCF's needs are met effectively with evidence-based treatment aligned with their individual health goals. The CF palliative care guideline highlights the importance of routine mental health screening throughout the lifespan, including at the start of new treatments that may impact mental and behavioral health [130]. Screening tools and best practice interventions beyond anxiety and depression for youth < 12 years of age are also available [131]. Given the impact of caregiver depression and anxiety on the health and functioning of the child, caregiver mental health screening is also recommended [39].

Although mental health screening is ideally completed in-person, screening remotely through telemedicine is preferable to no screening. Telemedicine was utilized within mental health care delivery prior to the pandemic, particularly for individuals with limited access to local mental health care [132–135]. Given the fact that the PHQ-9 includes a question regarding suicidality, clinical judgement should determine whether that question is appropriate via telemedicine [136]. For screening and delivery of mental health services, video capability as opposed to voice only, enhances the quality of the assessment and interaction.

3.6. Screening for health-related social needs (HRSN)/ social determinants of health (SDOH)

HRSN refer to "an individual's unmet, adverse social conditions (e.g. housing instability/ homelessness, food insecurity, unreliable access to medications, insurance, internet) that contribute to poor health and are a result of underlying social determinants of health" [137]. The impact of HRSN is substantial, with an estimated 80 % of health outcomes being accounted for by these factors [138–140]. Greater psychosocial risk has been associated with Medicaid insurance status and lower parental education [141]. Food insecurity, associated with poorer mental health for pwCF [142], has been reported in 30 % of pwCF, [143]. A comprehensive HRSN annual screening is suggested to identify factors amenable to intervention to promote optimal health and functioning in CF, including school [144] and vocational functioning. Many screening questions are part of the annual social work assessment. Screening alone is beneficial, even with incomplete resource referral or resolution [145]. HRSN resources can be found in the my.cff.org Resource Library. When HRSN are identified, more frequent follow-up with the CF team may be necessary (in person or via telemedicine) to monitor risk, support health, and ensure needs are being met (Table 1).

4. Conclusion

CF is a dynamic disease with a growing breadth and diversity of clinical needs and disease burden. The CF care model has proven successful over time with marked improvement in clinical outcomes, most notably survival. The introduction of CFTR modulator therapy has improved the health of many pwCF substantially; however, it is not

curative and long-term impact on disease progression remains to be seen. In addition, for many with CF, health stability is lacking, and frequent clinical assessment continues to be necessary, especially given additional health concerns with extended lifespans (osteoporosis, menopause, cancer, GI issues and more).

There are situations in which individualization of clinic visit frequency to every 4–6 months may be appropriate after careful consideration of a variety of health domains. Telemedicine and remote monitoring can complement in-person CF care delivery and should be integrated into the routine care model when possible, acknowledging their limitations. Empowered self-management and successful partnership between pwCF and their care teams are essential to the continued success of the CF care model. This guidance is based on currently available data and should evolve alongside new therapeutic advancements and knowledge over time. Data tracking the impact of changing care delivery on clinical outcomes and the experience of pwCF and their team will be critical to guide future evolution of the CF care model and ensure that access to high-quality specialized CF care continues to allow pwCF to live long and fulfilling lives.

Credit author's statement

DM Goetz, RF Brown were primary writers for the paper and DM Goetz was the corresponding author and completed all revisions, editing and submission.

SS Filigno was integral to the writing and concept of the paper and participated in editing and revisions.

SL Bichl was integral to the writing and concept of the paper and participated in editing and revisions.

AL Nelson was integral to the writing and concept of the paper and participated in editing and revisions.

CA Merlo, R Juel contributed to the writing of the paper and revisions.

P Lomas contributed to the concept and writing of the paper and revisions.

SE Hempstead contributed heavily to the writing of the paper and collated all references with revisions.

AW Brown, & PA Flume contributed heavily to the concept and writing of the paper and were integral to final revisions.

Financial disclosure

None of the authors have any financial relationships relevant to this article to disclose.

Funding

This work was supported by the CF Foundation.

Ethical approval

The study was exempt from Institutional Review Board approval.

Declaration of competing interest

The authors declare no conflict of interest.

Acknowledgements

The Care Model Position Paper Committee members included:
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