Canadian Cystic Fibrosis Conference

September 17-19, 2025

The guidelines are published, what next?

Implementation

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Cystic Fibrosis Foundation

CYSTIC FIBROSIS FOUNDATION®

Cystic Fibrosis Fibrose kystique Canada | w/o limits

Disclosures

Sarah Hempstead, MS

The following relationships exist:

Employee of the Cystic Fibrosis Foundation

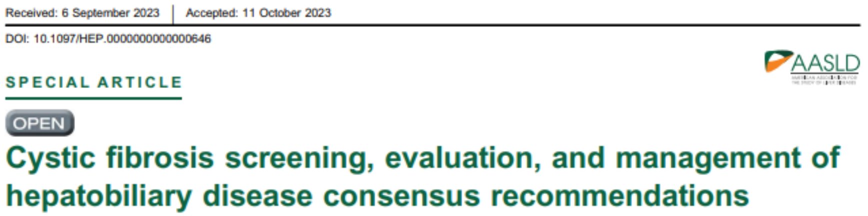
Overview

- Guidance Documents and Implementation Mechanisms
- Example: Depression and Anxiety
- What about the Care Model?

Types of Guidance Documents

Type of CFF Guidance Document	Panel of Experts	PICO questions	Systematic Literature Review	Grading of Evidence	Recommendation Statements 80% consensus	Public Comment	Publish in Peer Review Journal
Clinical Practice Guideline	**YES**	**YES**	**YES**	**YES**	**YES**	**YES**	**YES**
Consensus Guideline /Statement	**YES**	**YES**	**YES**	**NO**	**YES**	**YES**	**YES**
Technical Standards	YES	YES	No, Focused Review	No	YES	Focused	YES
Position Paper/ Guidance	**YES, Small panel**	**NO**	**No, Focused Review**	**NO**	**NO**	**YES**	**YES**
State of the Art	YES	NO	No, Focused Review	NO	NO	NO	YES
Clinical Considerations	YES, small panel	NO	NO	NO	NO	NO	NO, housed in my.CFF.org

Types of Guidance Documents



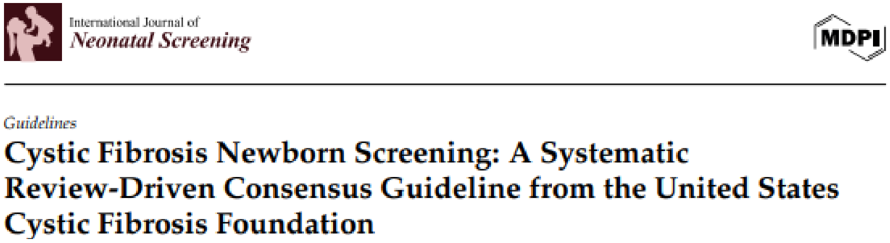
Received: 6 September 2023 | Accepted: 11 October 2023
DOI: 10.1097/HEP.0000000000000646

AASLD

SPECIAL ARTICLE

OPEN

Cystic fibrosis screening, evaluation, and management of hepatobiliary disease consensus recommendations

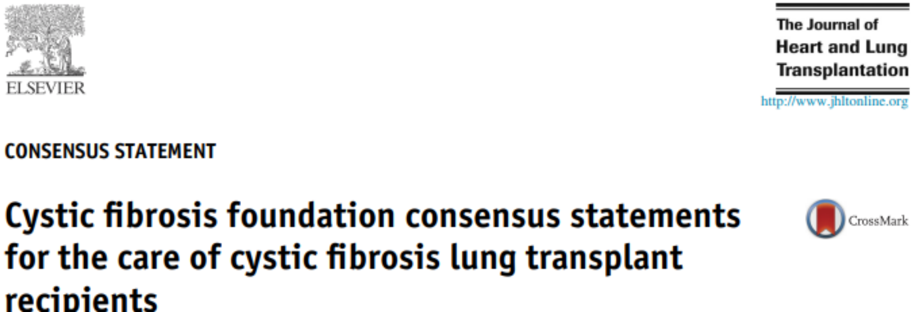


International Journal of *Neonatal Screening*

MDPI

Guidelines

Cystic Fibrosis Newborn Screening: A Systematic Review-Driven Consensus Guideline from the United States Cystic Fibrosis Foundation



ELSEVIER

The Journal of Heart and Lung Transplantation

http://www.jhltonline.org

CONSENSUS STATEMENT

CrossMark

Cystic fibrosis foundation consensus statements for the care of cystic fibrosis lung transplant recipients

Contents lists available at ScienceDirect

Journal of Cystic Fibrosis

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

ELSEVIER

Review

Cystic fibrosis foundation position paper: Redefining the cystic fibrosis care team

Contents lists available at ScienceDirect

Journal of Cystic Fibrosis

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

ELSEVIER

Original Article

Nutritional considerations for a new era: A CF foundation position paper

Contents lists available at ScienceDirect

Journal of Cystic Fibrosis

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

ELSEVIER

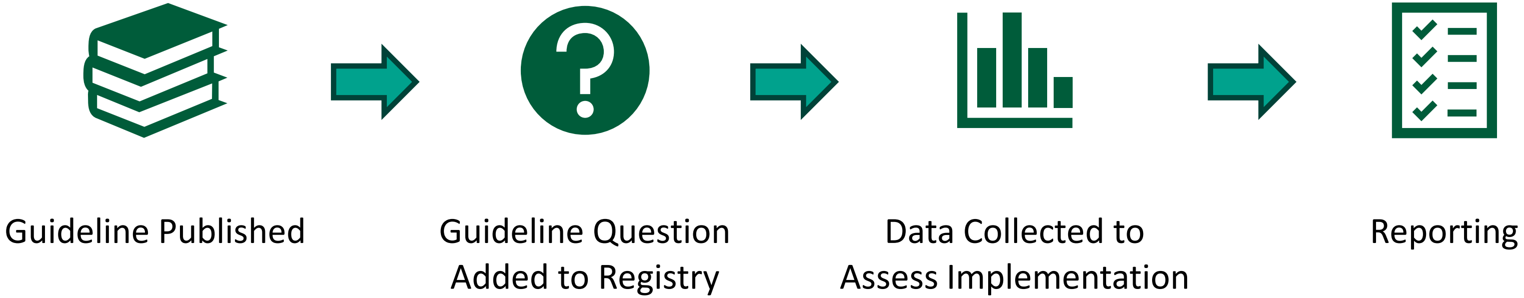
Original Article

Position paper: Models of post-transplant care for individuals with cystic fibrosis

Implementation Mechanisms

Patient Registry

Patient Registry and Guidelines



Guideline Published

Guideline Question Added to Registry

Data Collected to Assess Implementation

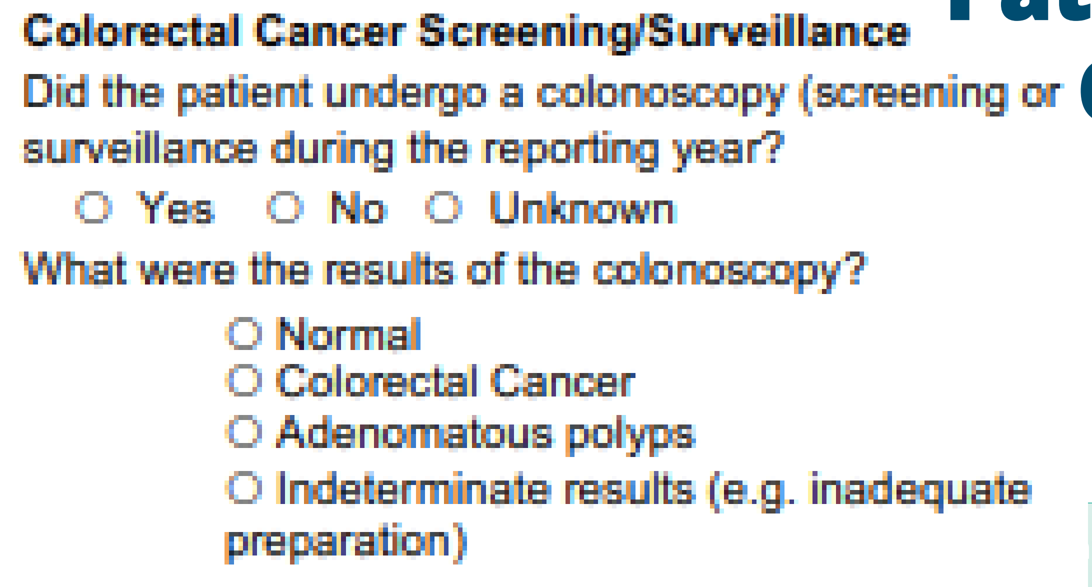
Reporting

Patient Registry Questions

- The CF Foundation recommends colonoscopy as the screening examination for CRC in individuals with CF
- The CF Foundation recommends that CRC screening begin at age 40 y in individuals with CF with continued rescreening every 5 y
- The CF Foundation recommends that individuals with CF who have undergone a colonoscopy that had any adenomatous polyps have surveillance colonoscopy in 3 y, unless shorter interval is indicated by individual findings, with subsequent intervals based on most recent endoscopic examination

Hadjiliadis D, Khoruts A, Zauber AG, Hempstead SE, Maisonneuve P, Lowenfels AB; Cystic Fibrosis Colorectal Cancer Screening Task Force. Cystic Fibrosis Colorectal Cancer Screening Consensus Recommendations. Gastroenterology. 2018 Feb;154(3):736-745.e14. doi: 10.1053/j.gastro.2017.12.012. Epub 2017 Dec 29.

Patient Registry Questions



Colorectal Cancer Screening/Surveillance

Did the patient undergo a colonoscopy (screening or surveillance during the reporting year?

○ Yes ○ No ○ Unknown

What were the results of the colonoscopy?

○ Normal
○ Colorectal Cancer
○ Adenomatous polyps
○ Indeterminate results (e.g. inadequate preparation)

- ation for CRC in individuals with CF
- The CF Foundation recommends that CRC screening begin at age 40 y in individuals with CF with continued rescreening every 5 y
- The CF Foundation recommends that individuals with CF who have undergone a colonoscopy that had any adenomatous polyps have surveillance colonoscopy in 3 y, unless shorter interval is indicated by individual findings, with subsequent intervals based on most recent endoscopic examination

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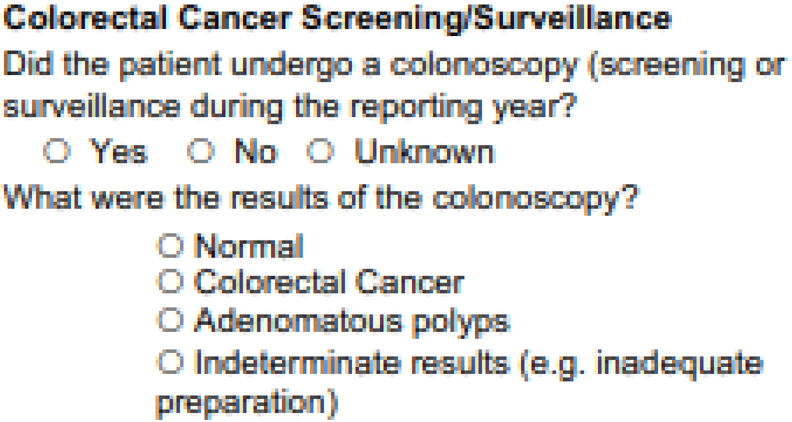
2024 Cystic Fibrosis Foundation Patient Registry Highlights Bethesda, Maryland ©2025 Cystic Fibrosis Foundation

Patient Registry Questions



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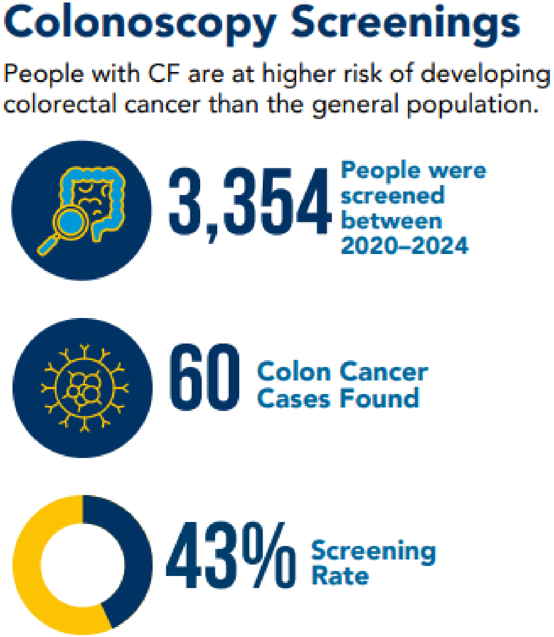


Colonoscopy Screenings

People with CF are at higher risk of developing colorectal cancer than the general population.

3,354 People were screened between 2020–2024

60 Colon Cancer Cases Found

43% Screening Rate

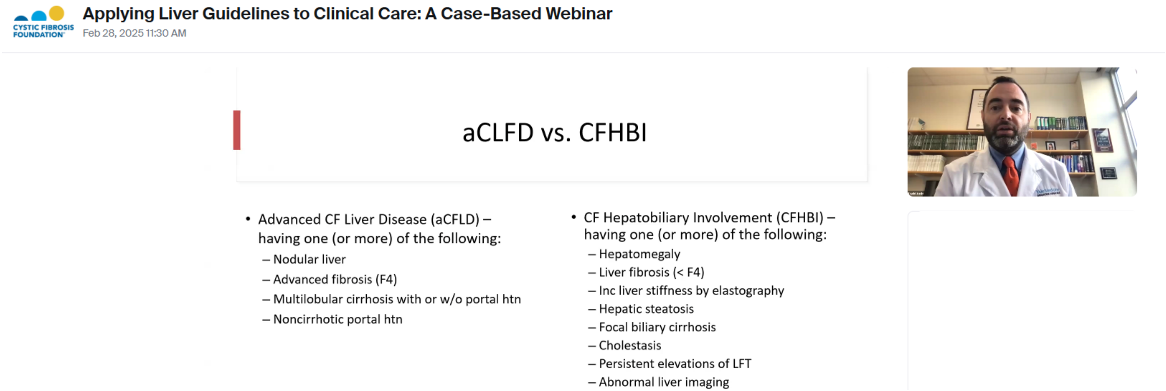
- The CF Foundation recommends colonoscopy as the screening examination for CRC in individuals with CF
- The CF Foundation recommends that CRC screening begin at age 40 y in individuals with CF with continued rescreening every 5 y
- The CF Foundation recommends that individuals with CF who have undergone a colonoscopy that had any adenomatous polyps have surveillance colonoscopy in 3 y, unless shorter interval is indicated by individual findings, with subsequent intervals based on most recent endoscopic examination

Hadjiliadis D, Khoruts A, Zauber AG, Hempstead SE, Maisonneuve P, Lowenfels AB; Cystic Fibrosis Colorectal Cancer Screening Task Force. Cystic Fibrosis Colorectal Cancer Screening Consensus Recommendations. Gastroenterology. 2018 Feb;154(3):736-745.e14. doi: 10.1053/j.gastro.2017.12.012. Epub 2017 Dec 29.

2024 Cystic Fibrosis Foundation Patient Registry Highlights
Bethesda, Maryland ©2025 Cystic Fibrosis Foundation

Education and Training

Webinars, Presentations, and Educational Materials

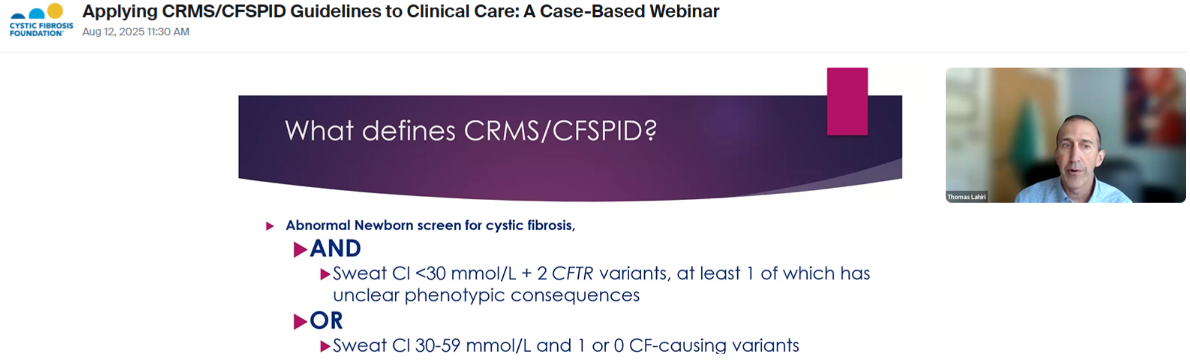


Applying Liver Guidelines to Clinical Care: A Case-Based Webinar

Feb 28, 2025 11:30 AM

aCLFD vs. CFHBI

- Advanced CF Liver Disease (aCFLD) – having one (or more) of the following:
  - Nodular liver
  - Advanced fibrosis (F4)
  - Multilobular cirrhosis with or w/o portal htn
  - Noncirrhotic portal htn
- CF Hepatobiliary Involvement (CFHBI) – having one (or more) of the following:
  - Hepatomegaly
  - Liver fibrosis (< F4)
  - Inc liver stiffness by elastography
  - Hepatic steatosis
  - Focal biliary cirrhosis
  - Cholestasis
  - Persistent elevations of LFT
  - Abnormal liver imaging



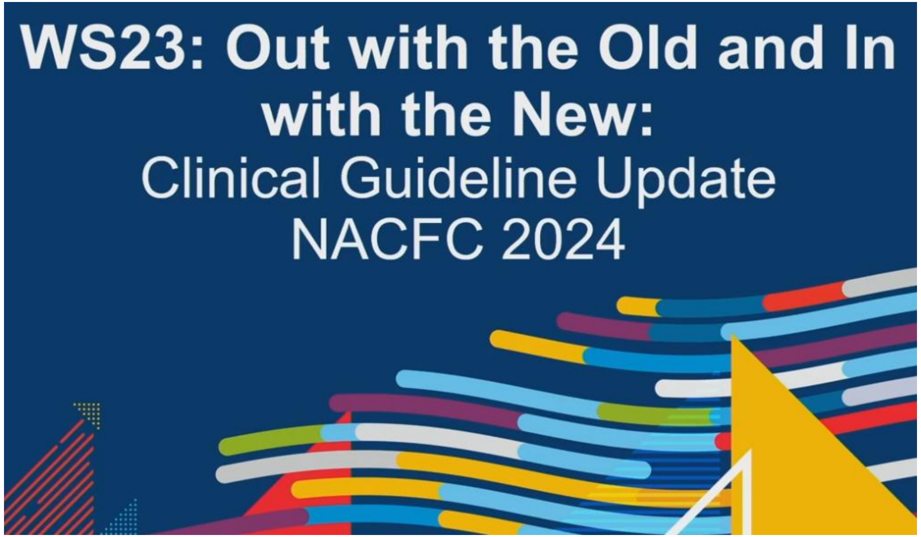
Applying CRMS/CFSPID Guidelines to Clinical Care: A Case-Based Webinar

Aug 12, 2025 11:30 AM

What defines CRMS/CFSPID?

- Abnormal Newborn screen for cystic fibrosis,
  - **AND**
    - Sweat Cl <30 mmol/L + 2 *CFTR* variants, at least 1 of which has unclear phenotypic consequences
  - **OR**
    - Sweat Cl 30-59 mmol/L and 1 or 0 CF-causing variants

Webinars, Presentations, and Educational Materials

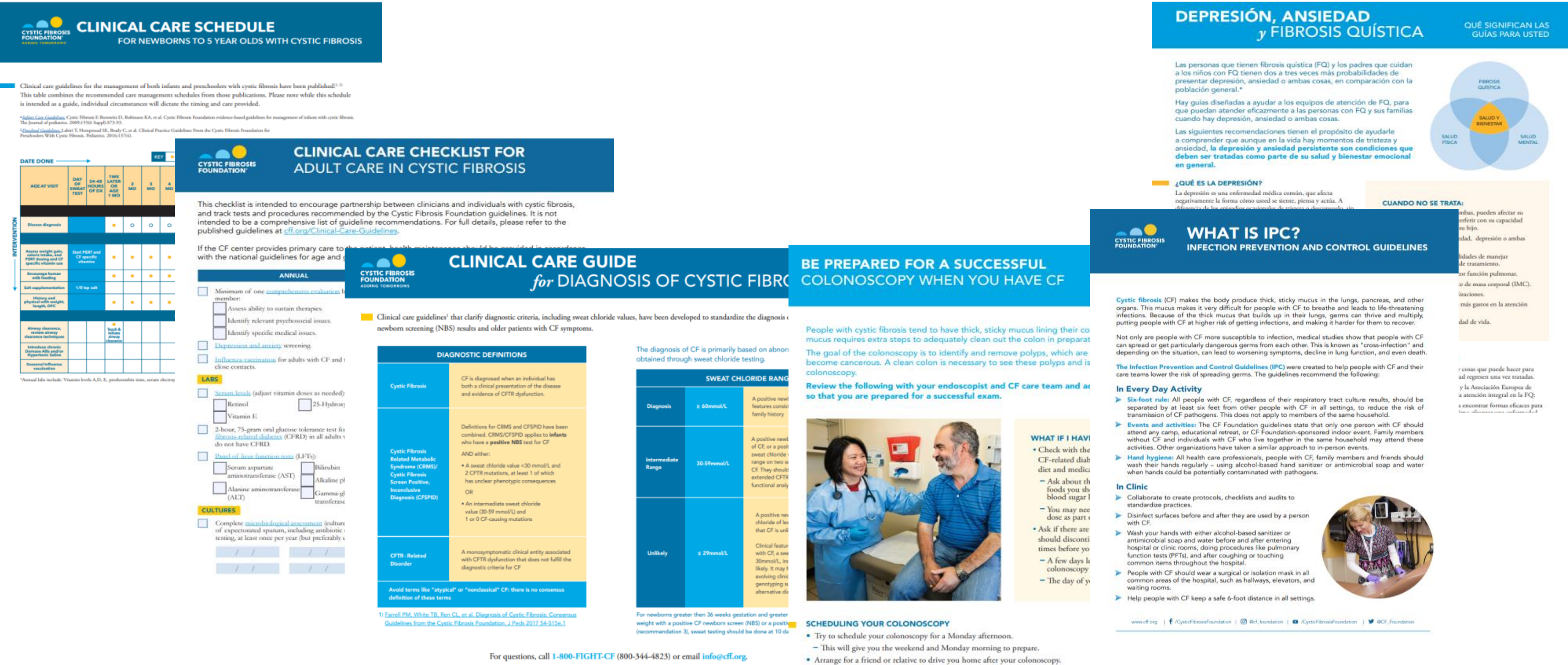


WS23: Out with the Old and In with the New:

Clinical Guideline Update

NACFC 2024

Webinars, Presentations, and Educational Materials



CYSTIC FIBROSIS FOUNDATION
ADDING TOMORROWS

CLINICAL CARE SCHEDULE
FOR NEWBORNS TO 5 YEAR OLDS WITH CYSTIC FIBROSIS

Clinical care guidelines for the management of both infants and preschoolers with cystic fibrosis have been published.[1, 2] This table combines the recommended care management schedules from those publications. Please note while this schedule is intended as a guide, individual circumstances will dictate the timing and care provided.

[1] Infant Care Guidelines: Cystic Fibrosis F, Borowitz D, Robinson KA, et al. Cystic Fibrosis Foundation evidence-based guidelines for management of infants with cystic fibrosis. The Journal of pediatrics. 2009;155(6 Suppl):S73-93.

[2] Preschool Guidelines: Lahiri T, Hempstead SE, Brady C, et al. Clinical Practice Guidelines From the Cystic Fibrosis Foundation for Preschoolers With Cystic Fibrosis. Pediatrics. 2016;137(4).

KEY ●

DATE DONE →

INTERVENTION

AGE AT VISIT	DAY OF SWEAT TEST	24-48 HOURS OF DX	1WK LATER OR AGE 1 MO	2 MO	3 MO	4 MO
Discuss diagnosis			●	○	○	○
Assess weight gain, caloric intake, and PERT dosing and CF specific vitamin use	Start PERT and CF specific vitamins		●	●	●	●
Encourage human milk feeding			●	●	●	●
Salt supplementation	1/8 tsp salt					
History and physical with weight, length, OFC			●	●	●	●
Airway clearance, review airway clearance techniques			● Teach & initiate airway clearance			
Introduce chronic Dornase Alfa and/or Hypertonic Saline						
Seasonal influenza vaccination						

*Annual labs include: Vitamin levels A,D, E, prothrombin time, serum electro

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CYSTIC FIBROSIS FOUNDATION

CLINICAL CARE CHECKLIST FOR ADULT CARE IN CYSTIC FIBROSIS

This checklist is intended to encourage partnership between clinicians and individuals with cystic fibrosis, and track tests and procedures recommended by the Cystic Fibrosis Foundation guidelines. It is not intended to be a comprehensive list of guideline recommendations. For full details, please refer to the published guidelines at cff.org/Clinical-Care-Guidelines.

If the CF center provides primary care to
with the national guidelines for age and

ANNUAL

- ☐ Minimum of one comprehensive evaluation b
member:
  - ☐ Assess ability to sustain therapies.
  - ☐ Identify relevant psychosocial issues.
  - ☐ Identify specific medical issues.
- ☐ Depression and anxiety screening.
- ☐ Influenza vaccination for adults with CF and
close contacts.

LABS

- ☐ Serum levels (adjust vitamin doses as needed):
  - ☐ Retinol
  - ☐ 25-Hydrox
  - ☐ Vitamin E
- ☐ 2-hour, 75-gram oral glucose tolerance test fo
fibrosis-related diabetes (CFRD) in all adults
do not have CFRD.
- ☐ Panel of liver function tests (LFTs):
  - ☐ Serum aspartate aminotransferase (AST)
  - ☐ Bilirubin
  - ☐ Alanine aminotransferase (ALT)
  - ☐ Alkaline p
  - ☐ Gamma-g
transferase

CULTURES

- ☐ Complete microbiological assessment (culture
of expectorated sputum, including antibiotic
testing, at least once per year (but preferably

/ /
/ /
/ /
/ /

CYSTIC FIBROSIS FOUNDATION
ADDING TOMORROWS

CLINICAL CARE GUIDE *for* DIAGNOSIS OF CYSTIC FIBRO

Clinical care guidelines[1] that clarify diagnostic criteria, including sweat chloride values, have been developed to standardize the diagnosis
newborn screening (NBS) results and older patients with CF symptoms.

DIAGNOSTIC DEFINITIONS	
Cystic Fibrosis	CF is diagnosed when an individual has both a clinical presentation of the disease and evidence of CFTR dysfunction.
Cystic Fibrosis Related Metabolic Syndrome (CRMS)/ Cystic Fibrosis Screen Positive, Inconclusive Diagnosis (CFSPID)	Definitions for CRMS and CFSPID have been combined. CRMS/CFSPID applies to **infants** who have a **positive NBS** test for CF AND either: • A sweat chloride value <30 mmol/L and 2 CFTR mutations, at least 1 of which has unclear phenotypic consequences OR • An intermediate sweat chloride value (30-59 mmol/L) and 1 or 0 CF-causing mutations
CFTR - Related Disorder	A monosymptomatic clinical entity associated with CFTR dysfunction that does not fulfill the diagnostic criteria for CF
Avoid terms like "atypical" or "nonclassical" CF: there is no consensus definition of these terms	

1) Farrell PM, White TB, Ren CL, et al. Diagnosis of Cystic Fibrosis: Consensus Guidelines from the Cystic Fibrosis Foundation. J Peds 2017 S4-S15e.1

The diagnosis of CF is primarily based on abnor
obtained through sweat chloride testing.

SWEAT CHLORIDE RANG		
Diagnosis	**≥ 60mmol/L**	A positive newb features consist family history
Intermediate Range	**30-59mmol/L**	A positive newb of CF, or a posit sweat chloride range on two s CF. They should extended CFTR functional analy
Unlikely	**≤ 29mmol/L**	A positive new chloride of les that CF is unl Clinical featur with CF, a swe 30mmol/L, inc likely. It may h evolving clinic genotyping su alternative dia

For newborns greater then 36 weeks gestation and greater
weight with a positive CF newborn screen (NBS) or a positi
(recommendation 3), sweat testing should be done at 10 da

For questions, call 1-800-FIGHT-CF (800-344-4823) or email info@cff.org.

BE PREPARED FOR A SUCCESSFUL COLONOSCOPY WHEN YOU HAVE CF

People with cystic fibrosis tend to have thick, sticky mucus lining their co
mucus requires extra steps to adequately clean out the colon in preparat

The goal of the colonoscopy is to identify and remove polyps, which are
become cancerous. A clean colon is necessary to see these polyps and is
colonoscopy.

**Review the following with your endoscopist and CF care team and a
so that you are prepared for a successful exam.**

WHAT IF I HAV

- Check with the
CF-related diab
diet and medica
  - Ask about th
foods you sh
blood sugar l
  - You may nee
dose as part
- Ask if there are
should disconti
times before yo
  - A few days l
colonoscopy
  - The day of y

SCHEDULING YOUR COLONOSCOPY

- Try to schedule your colonoscopy for a Monday afternoon.
  - This will give you the weekend and Monday morning to prepare.
- Arrange for a friend or relative to drive you home after your colonoscopy.

DEPRESIÓN, ANSIEDAD *y* FIBROSIS QUÍSTICA

QUÉ SIGNIFICAN LAS GUÍAS PARA USTED

Las personas que tienen fibrosis quística (FQ) y los padres que cuidan a los niños con FQ tienen dos a tres veces más probabilidades de presentar depresión, ansiedad o ambas cosas, en comparación con la población general.*

Hay guías diseñadas a ayudar a los equipos de atención de FQ, para que puedan atender eficazmente a las personas con FQ y sus familias cuando hay depresión, ansiedad o ambas cosas.

Las siguientes recomendaciones tienen el propósito de ayudarle a comprender que aunque en la vida hay momentos de tristeza y ansiedad, **la depresión y ansiedad persistente son condiciones que deben ser tratadas como parte de su salud y bienestar emocional en general.**

FIBROSIS QUÍSTICA
SALUD Y BIENESTAR
SALUD FÍSICA
SALUD MENTAL

¿QUÉ ES LA DEPRESIÓN?

La depresión es una enfermedad médica común, que afecta negativamente la forma cómo usted se siente, piensa y actúa. A

CUANDO NO SE TRATA:

mbas, pueden afectar su
erferir con su capacidad
u hijo.
edad, depresión o ambas
lidades de manejar
de tratamiento.
or función pulmonar.
e de masa corporal (IMC).
lizaciones.
más gastos en la atención
dad de vida.

cosas que puede hacer para
ad regresen una vez tratadas.
y la Asociación Europea de
a atención integral en la FQ:
a encontrar formas eficaces para

CYSTIC FIBROSIS FOUNDATION

WHAT IS IPC?
INFECTION PREVENTION AND CONTROL GUIDELINES

Cystic fibrosis (CF) makes the body produce thick, sticky mucus in the lungs, pancreas, and other organs. This mucus makes it very difficult for people with CF to breathe and leads to life-threatening infections. Because of the thick mucus that builds up in their lungs, germs can thrive and multiply, putting people with CF at higher risk of getting infections, and making it harder for them to recover.

Not only are people with CF more susceptible to infection, medical studies show that people with CF can spread or get particularly dangerous germs from each other. This is known as "cross-infection" and depending on the situation, can lead to worsening symptoms, decline in lung function, and even death.

The Infection Prevention and Control Guidelines (IPC) were created to help people with CF and their care teams lower the risk of spreading germs. The guidelines recommend the following:

In Every Day Activity

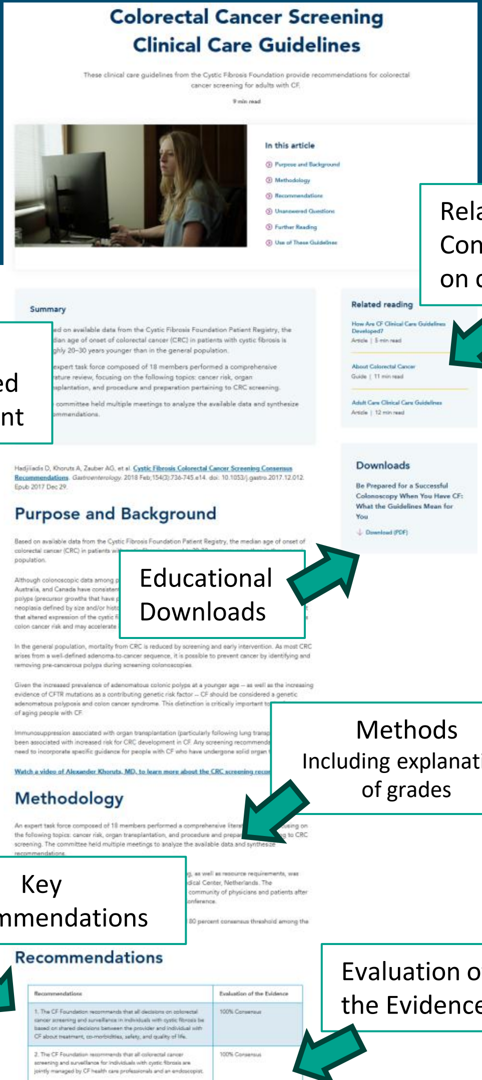
- ➤ **Six-foot rule:** All people with CF, regardless of their respiratory tract culture results, should be separated by at least six feet from other people with CF in all settings, to reduce the risk of transmission of CF pathogens. This does not apply to members of the same household.
- ➤ **Events and activities:** The CF Foundation guidelines state that only one person with CF should attend any camp, educational retreat, or CF Foundation-sponsored indoor event. Family members without CF and individuals with CF who live together in the same household may attend these activities. Other organizations have taken a similar approach to in-person events.
- ➤ **Hand hygiene:** All health care professionals, people with CF, family members and friends should wash their hands regularly – using alcohol-based hand sanitizer or antimicrobial soap and water when hands could be potentially contaminated with pathogens.

In Clinic

- ➤ Collaborate to create protocols, checklists and audits to standardize practices.
- ➤ Disinfect surfaces before and after they are used by a person with CF.
- ➤ Wash your hands with either alcohol-based sanitizer or antimicrobial soap and water before and after entering hospital or clinic rooms, doing procedures like pulmonary function tests (PFTs), and after coughing or touching common items throughout the hospital.
- ➤ People with CF should wear a surgical or isolation mask in all common areas of the hospital, such as hallways, elevators, and waiting rooms.
- ➤ Help people with CF keep a safe 6-foot distance in all settings.

www.cff.org | /CysticFibrosisFoundation | @cf_foundation | /CysticFibrosisFoundation | @CF_Foundation

Webinars, Presentations, and Educational Materials



Colorectal Cancer Screening Clinical Care Guidelines

These clinical care guidelines from the Cystic Fibrosis Foundation provide recommendations for colorectal cancer screening for adults with CF.

9 min read

In this article

- Purpose and Background
- Methodology
- Recommendations
- Unanswered Questions
- Further Reading
- Use of These Guidelines

Summary

…ed on available data from the Cystic Fibrosis Foundation Patient Registry, the …dian age of onset of colorectal cancer (CRC) in patients with cystic fibrosis is …ghly 20–30 years younger than in the general population.

…expert task force composed of 18 members performed a comprehensive …ature review, focusing on the following topics: cancer risk, organ …splantation, and procedure and preparation pertaining to CRC screening.

… committee held multiple meetings to analyze the available data and synthesize …mmendations.

Related reading

How Are CF Clinical Care Guidelines Developed?
Article | 5 min read

About Colorectal Cancer
Guide | 11 min read

Adult Care Clinical Care Guidelines
Article | 12 min read

Hadjiliadis D, Khoruts A, Zauber AG, et al. Cystic Fibrosis Colorectal Cancer Screening Consensus Recommendations. *Gastroenterology*. 2018 Feb;154(3):736-745.e14. doi: 10.1053/j.gastro.2017.12.012. Epub 2017 Dec 29.

Downloads

Be Prepared for a Successful Colonoscopy When You Have CF: What the Guidelines Mean for You

Download (PDF)

Purpose and Background

Based on available data from the Cystic Fibrosis Foundation Patient Registry, the median age of onset of colorectal cancer (CRC) in patients wi… population.

Although colonoscopic data among p… Australia, and Canada have consisten… polyps (precursor growths that have p… neoplasia defined by size and/or histo… that altered expression of the cystic fi… colon cancer risk and may accelerate…

In the general population, mortality from CRC is reduced by screening and early intervention. As most CRC arises from a well-defined adenoma-to-cancer sequence, it is possible to prevent cancer by identifying and removing pre-cancerous polyps during screening colonoscopies.

Given the increased prevalence of adenomatous colonic polyps at a younger age -- as well as the increasing evidence of CFTR mutations as a contributing genetic risk factor -- CF should be considered a genetic adenomatous polyposis and colon cancer syndrome. This distinction is critically important to… of aging people with CF.

Immunosuppression associated with organ transplantation (particularly following lung transp… been associated with increased risk for CRC development in CF. Any screening recommenda… need to incorporate specific guidance for people with CF who have undergone solid organ t…

Watch a video of Alexander Khoruts, MD, to learn more about the CRC screening recom…

Methodology

An expert task force composed of 18 members performed a comprehensive litera… …using on the following topics: cancer risk, organ transplantation, and procedure and prepar… …g to CRC screening. The committee held multiple meetings to analyze the available data and synthesize recommendations.

…g, as well as resource requirements, was …dical Center, Netherlands. The … community of physicians and patients after …onference.

… 80 percent consensus threshold among the

Recommendations

Recommendations	Evaluation of the Evidence
1. The CF Foundation recommends that all decisions on colorectal cancer screening and surveillance in individuals with cystic fibrosis be based on shared decisions between the provider and individual with CF about treatment, co-morbidities, safety, and quality of life.	100% Consensus
2. The CF Foundation recommends that all colorectal cancer screening and surveillance for individuals with cystic fibrosis are jointly managed by CF health care professionals and an endoscopist.	100% Consensus

Related Content on cff.org

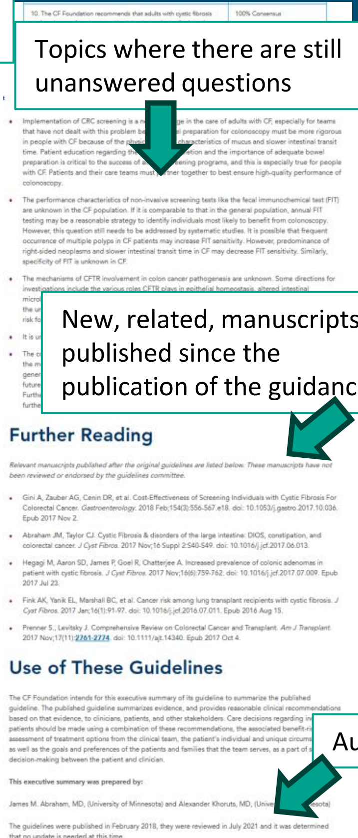
Link to published document

Educational Downloads

Methods
Including explanation of grades

Key Recommendations

Evaluation of the Evidence



10. The CF Foundation recommends that adults with cystic fibrosis	100% Consensus

- Implementation of CRC screening is a ne… …ge in the care of adults with CF, especially for teams that have not dealt with this problem be… …el preparation for colonoscopy must be more rigorous in people with CF because of the physio… …characteristics of mucus and slower intestinal transit time. Patient education regarding the… …etion and the importance of adequate bowel preparation is critical to the success of a… …eening programs, and this is especially true for people with CF. Patients and their care teams must p…rtner together to best ensure high-quality performance of colonoscopy.
- The performance characteristics of non-invasive screening tests like the fecal immunochemical test (FIT) are unknown in the CF population. If it is comparable to that in the general population, annual FIT testing may be a reasonable strategy to identify individuals most likely to benefit from colonoscopy. However, this question still needs to be addressed by systematic studies. It is possible that frequent occurrence of multiple polyps in CF patients may increase FIT sensitivity. However, predominance of right-sided neoplasms and slower intestinal transit time in CF may decrease FIT sensitivity. Similarly, specificity of FIT is unknown in CF.
- The mechanisms of CFTR involvement in colon cancer pathogenesis are unknown. Some directions for investigations include the various roles CFTR plays in epithelial homeostasis, altered intestinal microb… the un… risk fo…
- It is un…
- The c… the m… gener… future… Furthe… furthe…

Further Reading

Relevant manuscripts published after the original guidelines are listed below. These manuscripts have not been reviewed or endorsed by the guidelines committee.

- Gini A, Zauber AG, Cenin DR, et al. Cost-Effectiveness of Screening Individuals with Cystic Fibrosis For Colorectal Cancer. *Gastroenterology*. 2018 Feb;154(3):556-567.e18. doi: 10.1053/j.gastro.2017.10.036. Epub 2017 Nov 2.
- Abraham JM, Taylor CJ. Cystic Fibrosis & disorders of the large intestine: DIOS, constipation, and colorectal cancer. *J Cyst Fibros*. 2017 Nov;16 Suppl 2:S40-S49. doi: 10.1016/j.jcf.2017.06.013.
- Hegagi M, Aaron SD, James P, Goel R, Chatterjee A. Increased prevalence of colonic adenomas in patient with cystic fibrosis. *J Cyst Fibros*. 2017 Nov;16(6):759-762. doi: 10.1016/j.jcf.2017.07.009. Epub 2017 Jul 23.
- Fink AK, Yanik EL, Marshall BC, et al. Cancer risk among lung transplant recipients with cystic fibrosis. *J Cyst Fibros*. 2017 Jan;16(1):91-97. doi: 10.1016/j.jcf.2016.07.011. Epub 2016 Aug 15.
- Prenner S., Levitsky J. Comprehensive Review on Colorectal Cancer and Transplant. *Am J Transplant*. 2017 Nov;17(11):2761-2774. doi: 10.1111/ajt.14340. Epub 2017 Oct 4.

Use of These Guidelines

The CF Foundation intends for this executive summary of its guideline to summarize the published guideline. The published guideline summarizes evidence, and provides reasonable clinical recommendations based on that evidence, to clinicians, patients, and other stakeholders. Care decisions regarding in… patients should be made using a combination of these recommendations, the associated benefit-ri… assessment of treatment options from the clinical team, the patient's individual and unique circums… as well as the goals and preferences of the patients and families that the team serves, as a part of s… decision-making between the patient and clinician.

This executive summary was prepared by:

James M. Abraham, MD, (University of Minnesota) and Alexander Khoruts, MD, (Unive… …esota)

The guidelines were published in February 2018, they were reviewed in July 2021 and it was determined that no update is needed at this time.

Topics where there are still unanswered questions

New, related, manuscripts published since the publication of the guidance

Authors

About Colorectal Cancer

The risk for colorectal cancer in adults with cystic fibrosis is 5–10 times greater than the general population, and even higher for people with CF who receive a lung or other solid organ transplant. Find out how early screening can help reduce your risk.

11 min read

01. What Is Colorectal Cancer?

Colorectal cancer is the kind that begins in the colon or rectum. These cancers are also called colon cancer or rectal cancer, depending on where the cancer starts. Although many colorectal cancers can be prevented with regular colonoscopy screenings, it remains the third leading cause of cancer deaths in the U.S. among the general population.[1]

Continuing Medical Education

Cystic Fibrosis Online Continuing Education Program

- Course One: **Newborn Screening and Diagnosis UPDATED**
- Course Two: **Diagnosis and Management**—Birth through Pre-School Years **UPDATED**
- Course Four: **The Role of Sweat Testing in the Diagnosis of CF**
- Course Five: **Integrating Mental Health into CF Care**
- Course Six: **Advanced Cystic Fibrosis Lung Disease & Lung Transplant**
- Course Seven: **Endocrine Conditions and Co-Morbidities in CF**
- Course Eight: **Palliative Care in Cystic Fibrosis**
- Course Nine: **Adolescent & Young Adult Health Care NEW**
- Course 10 & 11: **Gene Therapy Trials in Cystic Fibrosis – What Clinicians Need to Know NEW**

Quality Improvement

CF Learning Network (CFLN)

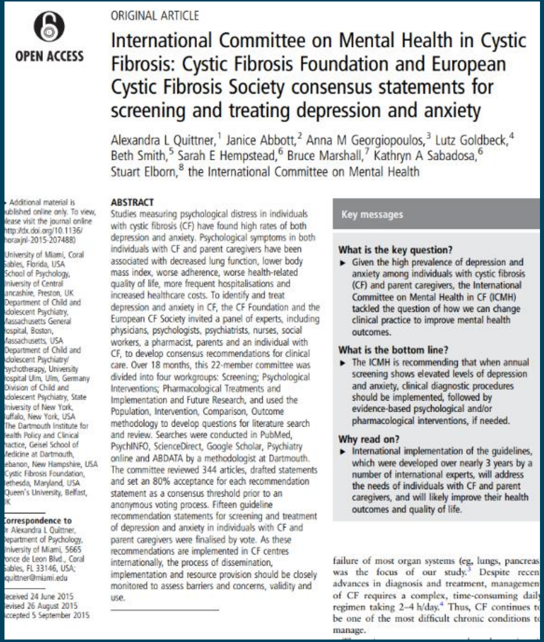
- Programs are invited to join the CFLN, currently 40 CF Programs participate

- Innovation Labs (iLab)
  - Topic selection is driven by the collaboration between CFF and the CFLN leadership team and not automatic for each guideline
  - Participating programs elect to to participate
  - Programs work on their own QI process to address the same question.
  - Programs track their own data to determine if the changes they are making make a difference. Aggregated data is used to track progress towards aims
  - Generation of Change Packages to help others implement

- Clinical care guidelines serve as the foundation for data collected at the network and lab level
- Implementing guidelines, or individual recommendation statements make good QI projects

- Virtual QI opportunities are open to all CF community members
  - Email cflearningnetwork@cff.org for questions

Example: Depression and Anxiety Guideline



OPEN ACCESS

ORIGINAL ARTICLE

International Committee on Mental Health in Cystic Fibrosis: Cystic Fibrosis Foundation and European Cystic Fibrosis Society consensus statements for screening and treating depression and anxiety

Alexandra L Quittner,[1] Janice Abbott,[2] Anna M Georgiopoulos,[3] Lutz Goldbeck,[4] Beth Smith,[5] Sarah E Hempstead,[6] Bruce Marshall,[7] Kathryn A Sabadosa,[6] Stuart Elborn,[8] the International Committee on Mental Health

Additional material is ublished online only. To view, lease visit the journal online http://dx.doi.org/10.1136/ horaxjnl-2015-207488)

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eceived 24 June 2015
evised 26 August 2015
ccepted 5 September 2015

ABSTRACT

Studies measuring psychological distress in individuals with cystic fibrosis (CF) have found high rates of both depression and anxiety. Psychological symptoms in both individuals with CF and parent caregivers have been associated with decreased lung function, lower body mass index, worse adherence, worse health-related quality of life, more frequent hospitalisations and increased healthcare costs. To identify and treat depression and anxiety in CF, the CF Foundation and the European CF Society invited a panel of experts, including physicians, psychologists, psychiatrists, nurses, social workers, a pharmacist, parents and an individual with CF, to develop consensus recommendations for clinical care. Over 18 months, this 22-member committee was divided into four workgroups: Screening; Psychological Interventions; Pharmacological Treatments and Implementation and Future Research, and used the Population, Intervention, Comparison, Outcome methodology to develop questions for literature search and review. Searches were conducted in PubMed, PsychINFO, ScienceDirect, Google Scholar, Psychiatry online and ABDATA by a methodologist at Dartmouth. The committee reviewed 344 articles, drafted statements and set an 80% acceptance for each recommendation statement as a consensus threshold prior to an anonymous voting process. Fifteen guideline recommendation statements for screening and treatment of depression and anxiety in individuals with CF and parent caregivers were finalised by vote. As these recommendations are implemented in CF centres internationally, the process of dissemination, implementation and resource provision should be closely monitored to assess barriers and concerns, validity and use.

Key messages

What is the key question?

- Given the high prevalence of depression and anxiety among individuals with cystic fibrosis (CF) and parent caregivers, the International Committee on Mental Health in CF (ICMH) tackled the question of how we can change clinical practice to improve mental health outcomes.

What is the bottom line?

- The ICMH is recommending that when annual screening shows elevated levels of depression and anxiety, clinical diagnostic procedures should be implemented, followed by evidence-based psychological and/or pharmacological interventions, if needed.

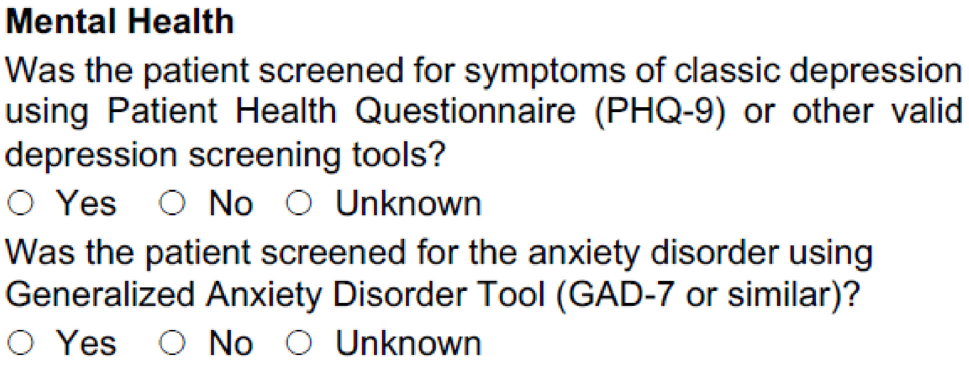
Why read on?

- International implementation of the guidelines, which were developed over nearly 3 years by a number of international experts, will address the needs of individuals with CF and parent caregivers, and will likely improve their health outcomes and quality of life.

failure of most organ systems (eg, lungs, pancreas was the focus of our study.[3] Despite recen advances in diagnosis and treatment, managemen of CF requires a complex, time-consuming dail regimen taking 2–4 h/day.[4] Thus, CF continues t be one of the most difficult chronic conditions t manage.

Cystic Fibrosis Foundation Patient Registry: Mental Health Screening:

Program Level Data Report



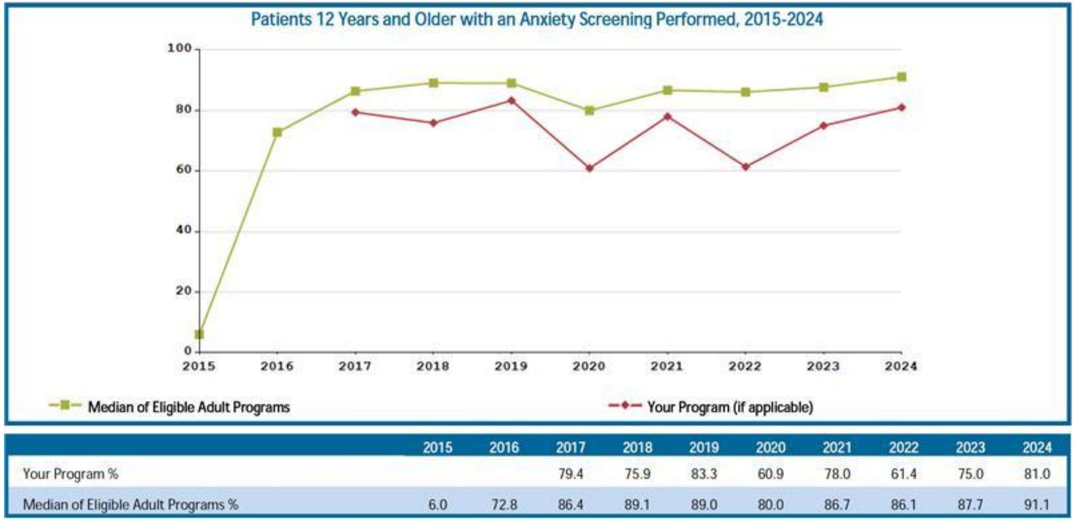
Mental Health

Was the patient screened for symptoms of classic depression using Patient Health Questionnaire (PHQ-9) or other valid depression screening tools?

○ Yes ○ No ○ Unknown

Was the patient screened for the anxiety disorder using Generalized Anxiety Disorder Tool (GAD-7 or similar)?

○ Yes ○ No ○ Unknown

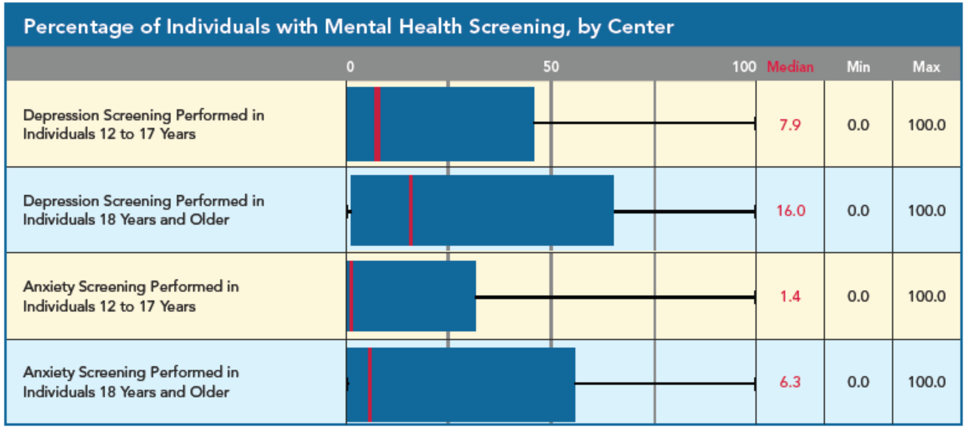


Patients 12 Years and Older with an Anxiety Screening Performed, 2015-2024

	2015	2016	2017	2018	2019	2020	2021	2022	2023	2024
Your Program %			79.4	75.9	83.3	60.9	78.0	61.4	75.0	81.0
Median of Eligible Adult Programs %	6.0	72.8	86.4	89.1	89.0	80.0	86.7	86.1	87.7	91.1

Cystic Fibrosis Foundation Patient Registry: Mental Health Screening

2015

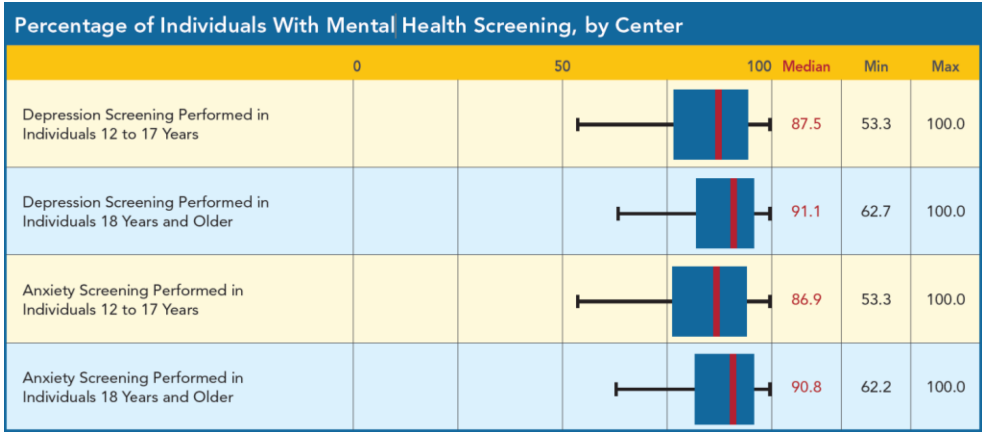


Percentage of Individuals with Mental Health Screening, by Center

	Median	Min	Max
Depression Screening Performed in Individuals 12 to 17 Years	7.9	0.0	100.0
Depression Screening Performed in Individuals 18 Years and Older	16.0	0.0	100.0
Anxiety Screening Performed in Individuals 12 to 17 Years	1.4	0.0	100.0
Anxiety Screening Performed in Individuals 18 Years and Older	6.3	0.0	100.0

Cystic Fibrosis Foundation Patient Registry
2015 Annual Data Report
Bethesda, Maryland

2024



Percentage of Individuals With Mental Health Screening, by Center

	Median	Min	Max
Depression Screening Performed in Individuals 12 to 17 Years	87.5	53.3	100.0
Depression Screening Performed in Individuals 18 Years and Older	91.1	62.7	100.0
Anxiety Screening Performed in Individuals 12 to 17 Years	86.9	53.3	100.0
Anxiety Screening Performed in Individuals 18 Years and Older	90.8	62.2	100.0

Cystic Fibrosis Foundation Patient Registry
2024 Annual Data Report
Bethesda, Maryland

Education and Training

Educational Materials

- Mental Health Advisory Committee: Education Workgroup
- Educational Content on CFF.org
- Mental Health Toolkit (dropbox)

Peer Support and Training

- Mental Health Advisory Committee: Peer Consultation
- Psychosocial Listserv
- Short Courses and Special Classes at NACFC
- CF Foundation Mentoring Program

To request access to the my.cff.org resource library please email mentalhealth@cff.org.

Cystic Fibrosis Online Education Program (CME)

Course Five: Integrating Mental Health into Cystic Fibrosis Care

Course Availability: 9/1/21–9/1/26

Audience: Members of adult and pediatric cystic fibrosis care team: cystic fibrosis clinical care coordinators, dietitians, fellows, genetic counselors, newborn screen coordinators, nurses, nurse practitioners, pharmacists, pharmacy technicians, physical therapists, physicians, physician assistants, psychologists, respiratory therapists, social workers.

CE Credits Provided: AMA PRA Category 1 Credits and ABP MOC 2 credit for Physicians. CE for Nursing (ANCC), Pharmacy (ACPE), and Social Work (IPLA). All other professions will be awarded non-physician attendance hours. Per their website, the American Psychological Association (APA) will accept credits awarded for this activity as the American Medical Association (AMA) is an approved organization of continuing education.

Course Duration: 4.0 credit hours

Enroll in course →

MODULE ONE
Depression and Anxiety in Adolescents and Adults with CF

MODULE TWO
Emotional Wellness in Children with CF under Age 12

MODULE THREE
Procedural Anxiety and Distress

MODULE FOUR
Substance Misuse

MODULE FIVE
Caregiver and Family Resilience

Now What? Quality Improvement

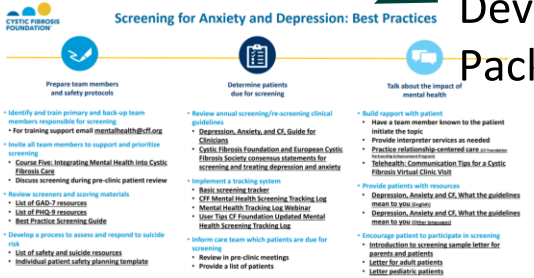
Cystic Fibrosis Learning Network (CFLN) Innovation Lab: Mental Health

Phase 1

Improve screening and re-screening rates

Complete

Develop Change Package



CYSTIC FIBROSIS FOUNDATION

Screening for Anxiety and Depression: Best Practices

Prepare team members and safety protocols

- Identify and train primary and back-up team members responsible for screening
  - For training support email mentalhealth@cff.org
- Invite all team members to support and prioritize screening
  - Course Five: Integrating Mental Health into Cystic Fibrosis Care
  - Discuss screening during pre-clinic patient review
- Review screeners and scoring materials
  - List of GAD-7 resources
  - List of PHQ-9 resources
  - Best Practice Screening Guide
- Develop a process to assess and respond to suicide risk
  - List of safety and suicide resources
  - Individual patient safety planning template

Determine patients due for screening

- Review annual screening/re-screening clinical guidelines
  - Depression, Anxiety, and CF, Guide for Clinicians
  - Cystic Fibrosis Foundation and European Cystic Fibrosis Society consensus statements for screening and treating depression and anxiety
- Implement a tracking system
  - Basic screening tracker
  - CFF Mental Health Screening Tracking Log
  - Mental Health Tracking Log Webinar
  - User Tips CF Foundation Updated Mental Health Screening Tracking Log
- Inform care team which patients are due for screening
  - Review in pre-clinic meetings
  - Provide a list of patients

Talk about the impact of mental health

- Build rapport with patient
  - Have a team member known to the patient initiate the topic
  - Provide interpreter services as needed
  - Practice relationship-centered care (CF Foundation Partnership Enhancement Program)
  - Telehealth: Communication Tips for a Cystic Fibrosis Virtual Clinic Visit
- Provide patients with resources
  - Depression, Anxiety and CF, What the guidelines mean to you (English)
  - Depression, Anxiety and CF, What the guidelines mean to you (Other languages)
- Encourage patient to participate in screening
  - Introduction to screening sample letter for parents and patients
  - Letter for adult patients
  - Letter pediatric patients

✚

Phase 2

Improve response to those with elevated scores

In progress

Develop Change Package

In progress

Publication of Results

Request access to my.cff.org resource library: mentalhealth@cff.org

Mental Health Research

PRIME

Prioritizing Research In Mental Health

Mental Health Advisory Committee
NACFC 2024

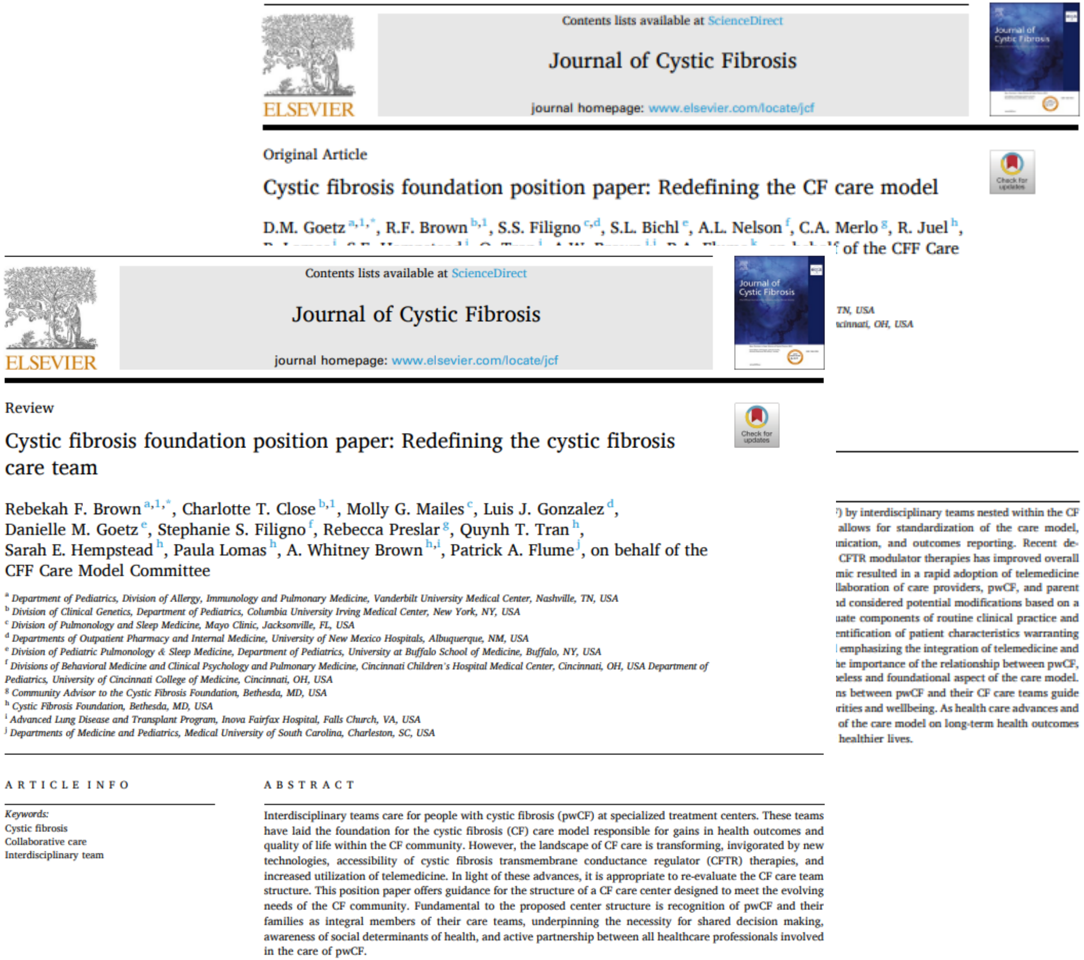
What about the Care Model?

Care Delivery
Care Teams

CYSTIC FIBROSIS
FOUNDATION®

Step 1:
Publish Position Papers

☑



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

D.M. Goetz [a,1,*], R.F. Brown [b,1], S.S. Filigno [c,d], S.L. Bichl [e], A.L. Nelson [f], C.A. Merlo [g], R. Juel [h],
f of the CFF Care

TN, USA
cinnati, OH, USA

) by interdisciplinary teams nested within the CF
allows for standardization of the care model,
nication, and outcomes reporting. Recent de-
CFTR modulator therapies has improved overall
mic resulted in a rapid adoption of telemedicine
llaboration of care providers, pwCF, and parent
d considered potential modifications based on a
uate components of routine clinical practice and
entification of patient characteristics warranting
emphasizing the integration of telemedicine and
he importance of the relationship between pwCF,
eless and foundational aspect of the care model.
ns between pwCF and their CF care teams guide
rities and wellbeing. As health care advances and
of the care model on long-term health outcomes
healthier lives.

Contents lists available at ScienceDirect

Journal of Cystic Fibrosis

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

Review

Cystic fibrosis foundation position paper: Redefining the cystic fibrosis care team

Rebekah F. Brown [a,1,*], Charlotte T. Close [b,1], Molly G. Mailes [c], Luis J. Gonzalez [d], Danielle M. Goetz [e], Stephanie S. Filigno [f], Rebecca Preslar [g], Quynh T. Tran [h], Sarah E. Hempstead [h], Paula Lomas [h], A. Whitney Brown [h,i], Patrick A. Flume [j], on behalf of the CFF Care Model Committee

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ARTICLE INFO

Keywords:
Cystic fibrosis
Collaborative care
Interdisciplinary team

ABSTRACT

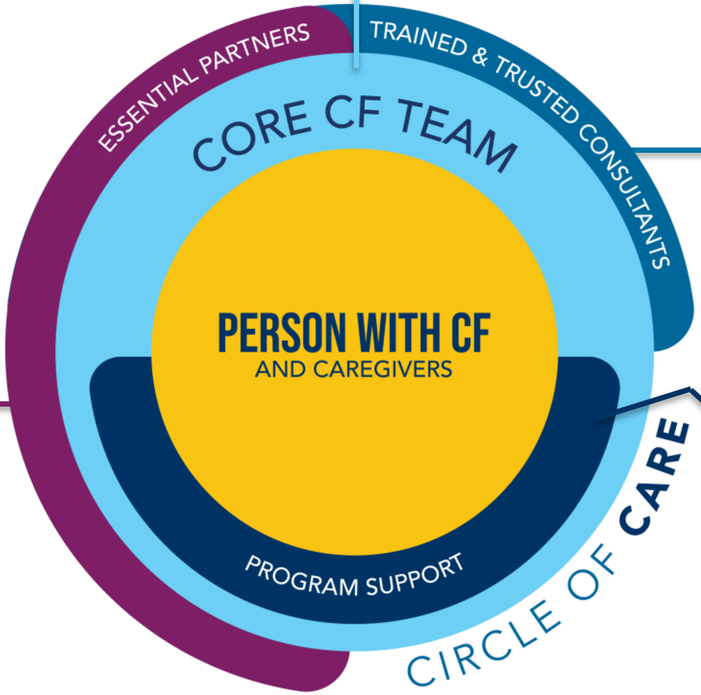
Interdisciplinary teams care for people with cystic fibrosis (pwCF) at specialized treatment centers. These teams have laid the foundation for the cystic fibrosis (CF) care model responsible for gains in health outcomes and quality of life within the CF community. However, the landscape of CF care is transforming, invigorated by new technologies, accessibility of cystic fibrosis transmembrane conductance regulator (CFTR) therapies, and increased utilization of telemedicine. In light of these advances, it is appropriate to re-evaluate the CF care team structure. This position paper offers guidance for the structure of a CF care center designed to meet the evolving needs of the CF community. Fundamental to the proposed center structure is recognition of pwCF and their families as integral members of their care teams, underpinning the necessity for shared decision making, awareness of social determinants of health, and active partnership between all healthcare professionals involved in the care of pwCF.

Step 2: Accreditation

Accreditation Standards	Center Committee is made up of CF Program Directors
	Standards are minimum requirements for programs to be accredited as part of the Care Center Network
Position Papers	Guidance on suggested best practices based on the state of the published literature at the time of publication

Step 3: Workforce Development and QI

Provides services required for routine clinical care that supports the health and well-being of all people with CF.



TRAINED & TRUSTED CONSULTANTS

Provide services to help manage less common CF and non-CF related complications.

ESSENTIAL PARTNERS

Have expertise in CF and work closely with the Core CF Team to manage CF-related complications that many, but not all, people with CF experience. May be part of Core CF Team at some programs.

PROGRAM SUPPORT

Offer services to support the Core CF Team but may not be directly involved in patient care.

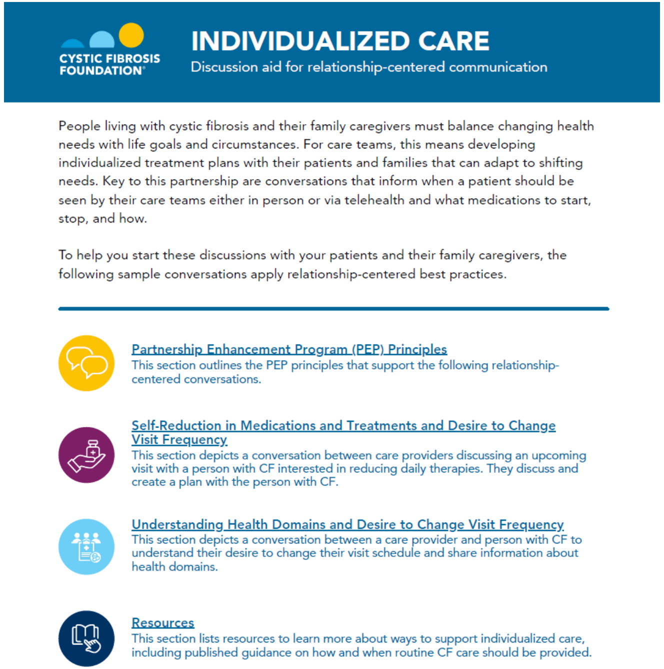
Step 3: Workforce Development and QI

Tools for Remote Care

Topic	Tools for Remote Care Study Title	Lead Investigators/Teams	Peds/Adult
Home Spirometry	**ICUPS:** Increasing the use of home spirometry and integrating it into clinic workflow	**Clement Ren – CHOP** Daniel Weiner – University of Pittsburgh Don Sanders – Indiana University	Peds
Home Spirometry	**ISPY:** Assessing accuracy of home spirometry and feasibility and acceptability for patients and clinics	**Thida Ong – Seattle Children's** Andrea Hartzler – UW Jaclyn Davis – Boston Children's Cori Daines – University of Arizona Rebekah Brown – Vanderbilt	Peds
Acute Sick Visits	**EUROPA:** Utilizing telehealth visits and remote testing to expedite and optimize clinical assessment for sick patients	**Noah Lechtzin – JHU** Randy Hunt – Emory Caralee Forseen, Kathleen McKie – Augusta Gina Hong – UPenn	Both
Home Cultures	**S2wAB-IT:** Assessing accuracy and reliability of self-obtained respiratory cultures, and implementation of home cultures as part of clinical care	**Chris Siracusa – Cincinnati Children's** **Jordana Hoppe – Children's Colorado** Elliott Dasenbrook – Cleveland Clinic Marianne Muhlebach – UNC Chapel Hill M. Veronica Indihar – U of Cincinnati Kathryn Moffett - WVU	Both
Remote Testing	**CF@HOME:** Utilizing an app to coordinate and encourage self-monitoring of health and performance of complication screening tests	**Scott Donaldson – UNC Chapel Hill** Jonathan Zuckerman – Maine Medical Center Alex Gifford – UHH Cleveland Adupa Rao - USC	Adult
Social Determinants of Health Screening	**ELICIT:** Improving Access to Care Through Remote Social Determinants of Health Screening and Intervention	**Bryan Garcia – UAB** **Didi Jennings - UVA** Greg Sawicki – Boston Children's Mike Powers - OSHU Dana Albon – UVA	Both
Frequency of Visits	**CF CARE MODEL:** The Cystic Fibrosis Collaborative Adjustments to Routine Clinical Examinations via Modification of Duration to the next Encounter	**David Finklea – UT Southwestern** Randy Hunt – Emory Christian Merlo –Johns Hopkins	Adult

Step 4: Education and Training

A Work in Progress



CYSTIC FIBROSIS FOUNDATION®

INDIVIDUALIZED CARE

Discussion aid for relationship-centered communication

People living with cystic fibrosis and their family caregivers must balance changing health needs with life goals and circumstances. For care teams, this means developing individualized treatment plans with their patients and families that can adapt to shifting needs. Key to this partnership are conversations that inform when a patient should be seen by their care teams either in person or via telehealth and what medications to start, stop, and how.

To help you start these discussions with your patients and their family caregivers, the following sample conversations apply relationship-centered best practices.

Partnership Enhancement Program (PEP) Principles
This section outlines the PEP principles that support the following relationship-centered conversations.

Self-Reduction in Medications and Treatments and Desire to Change Visit Frequency
This section depicts a conversation between care providers discussing an upcoming visit with a person with CF interested in reducing daily therapies. They discuss and create a plan with the person with CF.

Understanding Health Domains and Desire to Change Visit Frequency
This section depicts a conversation between a care provider and person with CF to understand their desire to change their visit schedule and share information about health domains.

Resources
This section lists resources to learn more about ways to support individualized care, including published guidance on how and when routine CF care should be provided.

CYSTIC FIBROSIS FOUNDATION®

When Should You Come To Clinic?

Things to Think About When Scheduling Your Next Visit

Quarterly visits to a CF Foundation-accredited care center are the foundation of your treatment plan. By partnering with your care team, you can discuss when your next routine in-person clinic visit should be scheduled.

If you are feeling well with stable health, you may be able to wait 4–6 months before your next clinic visit instead of every three months. This may be true especially if you stay in touch with your CF care team using telehealth, phone calls, or online check-ins. But if your health is changing, being examined in CF clinic sooner may help you get your health back on track. In discussing when to schedule your next clinic visit, consider the following.

How to Know if Your Health is Changing

LUNGS

Are you having…

- More coughing, chest tightness, or other breathing issues?
- A drop in lung function?
- A recent lung infection or flare-up (exacerbation)?
- Changes in your treatments like starting or stopping modulators or other medications?
- Trouble with physical activity or ability to exercise?

DIGESTIVE SYSTEM (GASTROINTESTINAL OR GI)

Are you noticing…

- Sudden changes in digestion (e.g., constipation, diarrhea) or other stomach issues?
- Changes to GI medications like enzymes or acid blockers?
- Abnormal liver test results?

ENDOCRINE SYSTEM (GLANDS THAT RELEASE HORMONES)

Are you…

- Recently diagnosed with CF-related diabetes (CFRD)?
- Having trouble controlling your CFRD?

NUTRITION

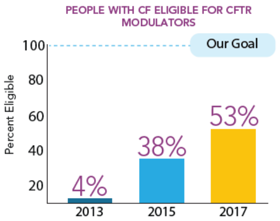
Are you…

- Losing or gaining 5% of your total body weight?
- Becoming underweight or overweight?
- Changing your diet or eating habits, including vitamins and supplements?
- Worried about your weight?

Implementation Opportunities and Challenges: Changing Healthcare Landscape

2017

CFTR MODULATORS



LUMACAFTOR/ IVACAFTOR

10,669 people with CF are eligible to take lumacaftor/ivacaftor

63% of people aged 6 or older with an FDA approved CFTR mutation are on lumacaftor/ ivacaftor

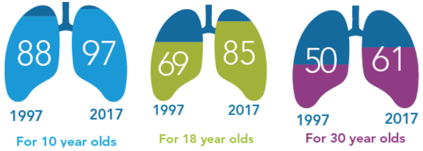
IVACAFTOR

4,293 people with CF are eligible to take ivacaftor

59% of people aged 2 or older with an FDA approved CFTR mutation are on ivacaftor

22,535 Hospitalizations

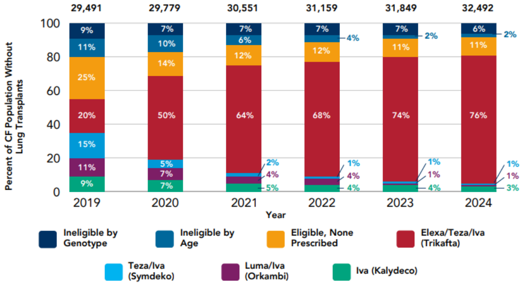
Median FEV$_1$ Percent Predicted



Median Predicted Survival

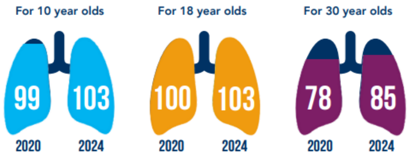
44 YEARS 2013-2017

2024



10,407 Hospitalizations

MEDIAN FEV$_1$ PERCENT PREDICTED



Median Predicted Survival

65 YEARS 2020–2024

2017 and 2024 CF Foundation Patient Registry Highlights Reports

Thank You.

Contact: shempstead@cff.org

Resources

Access my.cff.org
Email: mentalhealth@cff.org

Guideline or Guidance Document Questions:
Email: shempstead@cff.org

Quality Improvement
Email: cflearningnetwork@cff.org

CF Foundation Patient Registry
Email: Reghelp@cff.org

Indiana University CME