CYSTIC FIBROSIS
FOUNDATION®

2023

ADVANCED LUNG DISEASE
LUNG TRANSPLANT

Year in Review

ADVANCED LUNG DISEASE PROGRAM

Community ● Partnership ● Support

The Advanced CF Lung Disease (ACFLD) and Lung Transplant team is proud to share these updates from 2023 to members of the community, clinicians, researchers, and beyond. The Advanced Lung Disease and Transplant Initiative underscores that "No one left behind" is not a motto or slogan, but it is our very essence as a Foundation and community.

This effort aims to improve outcomes for individuals living with ACFLD including those pursuing, undergoing, or having experienced lung transplantation through quality improvement, guideline development, and research.

The CF Foundation has honored this commitment by funding $70 million in programs focused on optimizing lung transplant outcomes and prevention of chronic lung allograft dysfunction (CLAD) since 2017.

CARE | CURE
COMMUNITY

ADVANCED LUNG DISEASE (ALD)

- Advocacy
- Guideline Development
- ALD Registry
- Quality Improvement
- Toolkits

PALLIATIVE CARE

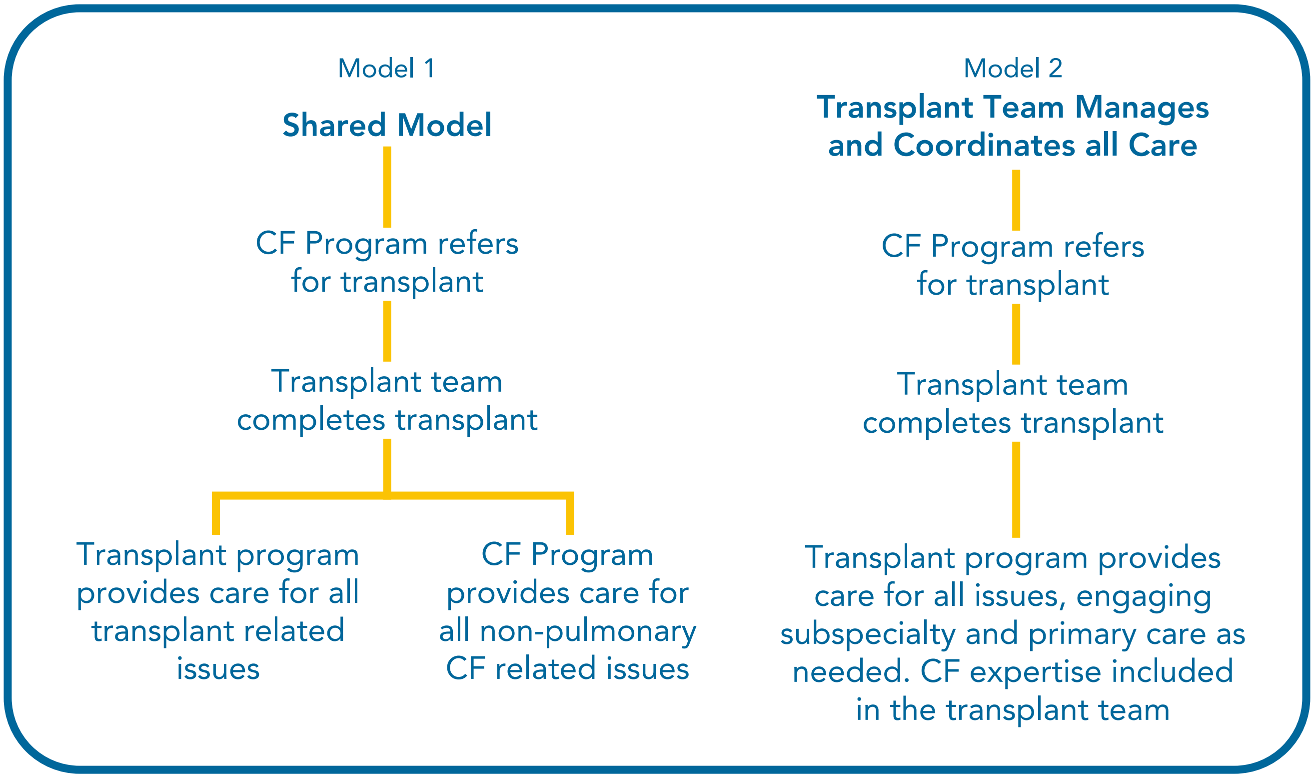
- Research
- Guideline Development
- Quality Improvement

LUNG TRANSPLANT

- Bronchial Brushings Repository
- Guideline Development
- Biorepository & Post-Tx Registry
- Quality Improvement
- Basic & Clinical Research
- Organ Allocation & Donor Availability
- Partnering with Industry

POST-LUNG TRANSPLANT MODELS OF CARE

On March 5, 2023, the *Journal of Cystic Fibrosis* published, *Position paper: Models of post-transplant care for individuals with CF*, highlighting two potential models for optimal care delivery for individuals with CF who are post-lung transplant.



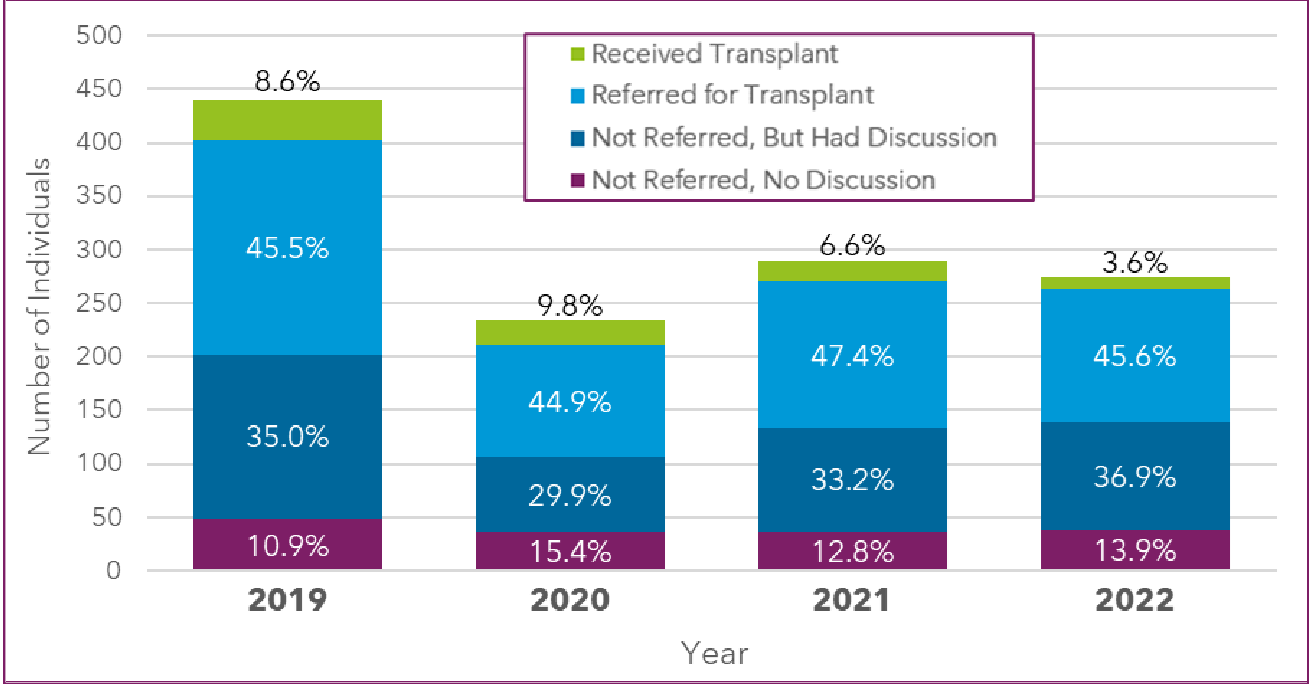
The first model's focus is on shared care between the CF and transplant teams, relying on excellent communication between the CF care and transplant teams. The second model utilizes the transplant team as care coordinators of the whole person.
This model emphasizes the need for knowledgeable, multidisciplinary clinicians on the transplant team who have expertise in managing CF or have access to specialists within the institution.

> *"Coordinating care between lung transplant and CF teams or consolidating care in one center that has expertise in both lung transplant and CF, is crucial to identifying complications and addressing them early to make the most of the gift of new lungs. The Guidelines of routine post-transplant care and modes of shared care provide a blueprint for best care after transplant."*
>
> **Joe Pilewski, MD**
> **University of Pittsburgh Medical Center**

2022 PATIENT REGISTRY ANNUAL DATA REPORT

The CFF Patient Registry began collecting data on advanced lung disease-specific case report forms in 2019. This data has been compiled and is now available in a dedicated chapter within the *2022 Patient Registry Annual Data Report*.

Referral Status for Advanced CF Lung Disease Individuals with FEV_1<30, 2019- 2022



- The above bar graph highlights the severity of disease at the time of transplant, referral for transplant, having discussions around transplant, and those who have not been referred or have recorded discussions with their care teams.
- About 50% of people eligible for transplant referral in 2022 were not referred. This echos the importance of our continued efforts to implement existing care guidelines for this group and encourage earlier conversations about lung transplant to optimize referral.

FUTURE RESEARCH!

We are excited to partner with the CF Patient Registry research team in the coming years to further study this data and ultimately open up requests for external investigators to do meaningful analyses.

UNDERSTANDING CHRONIC LUNG ALLOGRAFT DYSFUNCTION (CLAD)

2023 CLAD WORKSHOP

In December 2023, the CF foundation brought together experts in the field of CLAD from all across the U.S., Canada, and Europe to share current data and identify gaps in the field that still need to be addressed. This was a very productive meeting, resulting in the convening of a CLAD working group who will help inform and strategize our CLAD-related efforts, such as requests for applications (RFAs) and other funding opportunities to support much-needed clinical research in this field.

WHY CONTINUE TO FOCUS TRANSPLANT RESEARCH EFFORTS ON CLAD?

CLAD remains the major cause of death beyond two years after lung transplantation and develops in 50% of all patients by five years post-transplant.

An ongoing priority of this initiative is to identify therapies to detect, prevent, monitor, and treat CLAD.

CYSTIC FIBROSIS LUNG TRANSPLANT CONSORTIUM (CFLTC) BIOREPOSITORY AND REGISTRY

Launched in May 2021 with 15 transplant centers, this biorepository continues to collect biological samples from CF and non-CF lung transplant recipients and those awaiting a lung transplant.

At the end of 2023, over 800 individuals were enrolled and recruitment is ongoing.

Over 400 of these participants have CF

In the future, investigators will use these samples linked to clinical data to study the development and treatment of CLAD.

RESEARCH GRANTS

The CF Foundation is committed to identifying gaps in research in the field of lung transplant and funding studies that can have impact on individuals with CF who undergo transplant.

Highlights of 2023 Awarded Grants

- **Gliptin Use and Impact on CLAD**
  - Assessing the use of a common diabetes drug in the prevention of CLAD
- **Planning Grant: Allo-islet Cell Transplantation in Lung Transplant Recipients with Insulin Resistance**
  - Supporting early-stage protocol design and feasibility of islet cell transplantation to treat CF-related diabetes (CFRD)
- **Planning Grant: Randomized Control Trial of Azithromycin to Prevent CLAD**
  - Designing a multi-center randomized controlled trial of azithromycin to prevent CLAD after lung transplantation

Ongoing Funded Efforts

- 2021 CLAD RFA *(final year)*
- 2022 CTOT RFA *(final year)*
- CFLTC Biorepository and Registry Study
- CFTC Airway Brushings in CLAD Ancillary Study

Anticipated 2024 Grants

- New CLAD RFA
- New CTOT RFA

COMMUNITY EVENTS IN PARTNERSHIP WITH OTHER CF FOUNDATION PROGRAMS

Assisting people with CF, their families, and their caregivers throughout the transplant journey

RESEARCHCON

NOW YOU TELL ME: NAVIGATING CARE THROUGHOUT THE LUNG TRANSPLANT JOURNEY

A lung transplant expert, a person with CF, and a daughter who served as a caregiver throughout her mother's lung transplant journey speak on updated guidelines and unexpected realities.

LOSING YOUR BREATH, GAINING YOUR RESILIENCE

Keynote speaker, Ella Balasa, shares pivotal moments in her CF journey.

TEN AND 26–YRS POST-TRANSPLANT: WHAT I WISH I HAD KNOWN

Panel discussion with two adults with CF on their post- transplant experiences

NEXTBREATH

ADVOCATE, EMPOWER, AND THRIVE: NAVIGATING THE WORLD OF POST-TRANSPLANT

Co-chair Ashley Locke and her transplant physician, Dr. Sagana, answer questions related to transplant.

WINDED AND IMPATIENTLY WAITING: THE MANY JOURNEYS OF ADVANCED CF LUNG DISEASE

Two adults describe their experience with ACFLD, being listed for transplant, and finally receiving their transplant.

COMMUNITY INVOLVEMENT

PARTNERING WITH CF COMMUNITY MEMBERS REMAINS A PRIORITY

The Cystic Fibrosis Lung Transplant Consortium Steering Committee has welcomed two new community members to join in 2024, resulting in a total of four community members participating as active voices in this committee. Our community members are vital partners in understanding the needs of the CF lung transplant community and help to guide the priorities of our program.

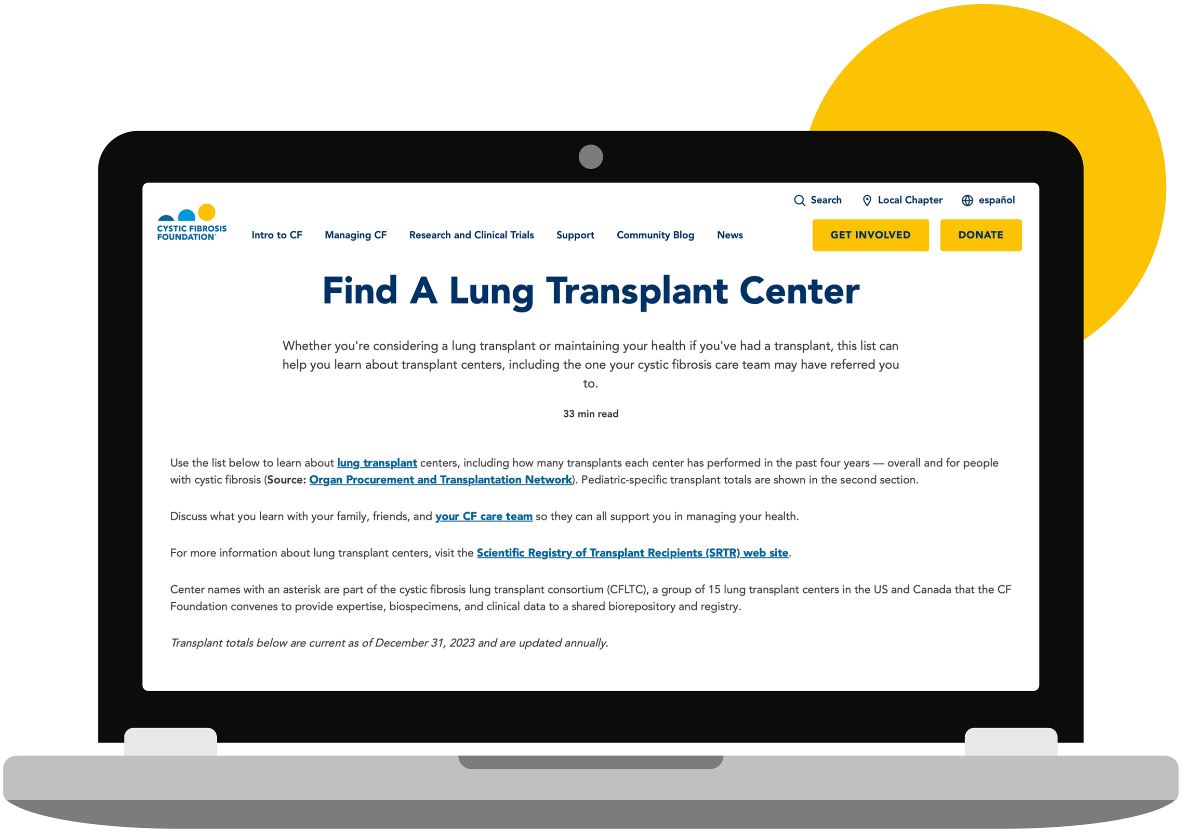
A BIG THANK YOU TO:

FANNY VLAHOS **IAN TISDALE**

APRIL BIGGS **SAMANTHA RICK**

COMMUNITY RESOURCES

For those considering a lung transplant or wanting to maintain their care post-transplant, check out the *Find a Lung Transplant Center* search tool on the CF Foundation's website.



Find A Lung Transplant Center

Whether you're considering a lung transplant or maintaining your health if you've had a transplant, this list can help you learn about transplant centers, including the one your cystic fibrosis care team may have referred you to.

33 min read

Use the list below to learn about lung transplant centers, including how many transplants each center has performed in the past four years — overall and for people with cystic fibrosis (**Source:** Organ Procurement and Transplantation Network). Pediatric-specific transplant totals are shown in the second section.

Discuss what you learn with your family, friends, and your CF care team so they can all support you in managing your health.

For more information about lung transplant centers, visit the Scientific Registry of Transplant Recipients (SRTR) web site.

Center names with an asterisk are part of the cystic fibrosis lung transplant consortium (CFLTC), a group of 15 lung transplant centers in the US and Canada that the CF Foundation convenes to provide expertise, biospecimens, and clinical data to a shared biorepository and registry.

Transplant totals below are current as of December 31, 2023 and are updated annually.

ACKNOWLEDGMENTS

RESOURCES

Winter 2018 Lung Transplant Initiative Update Paper

Fall 2020 Lung Transplant Initiative Update Paper

Summer 2022 Lung Transplant Initiative Update Paper

Characterization of chronic lung allograft dysfunction phenotypes using spectral and intrabreath oscillometry

Clinical Microsystem About Us

Find a Lung Transplant Center

Modify Lung Allocation by Blood Type

NextBreath 2023

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

Register to be an organ, eye, and tissue donor

ResearchCon 2023

Research to Cure, Models to Care: Advanced Lung Disease in Cystic Fibrosis

CONTACT THE TEAM: LUNGTRANSPLANT@CFF.ORG

CLINICAL INQUIRIES

Erin Tallarico,
Senior Director

Abigail Boyle,
Program Specialist

RESEARCH INQUIRIES

Abigail Thaxton,
Senior Manager

Bethelhem Markos,
Program Specialist

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