

Annual 2022 North American Cystic Fibrosis Conference Review

Leanne Saville, Pharm D
Sam Snow RDN, LRD
Christina Bond, LMSW
Deb Fueller, FNPC

Leanne Saville, PharmD



CF Pharmacist: What I Learned at NACFC

- Maternal and fetal outcomes in the era of CFTR modulators
- Vitamins/supplements and their impact on CF patients
- Immunization recommendations by the CF team
- Antibiotics for multidrug-resistant organisms (MDROs)

Maternal and Fetal Outcomes in the Era of CFTR Modulators

- Pregnancy has nearly doubled in the past 5 years
- CFTR modulators in pregnancy
 - Increases lung function
 - Increases cervical mucus pH
 - Improves nutritional status

Immunizations

- CF team can improve immunization rates with administering vaccinations at clinic visit
- Pharmacist/Pharmacy technician to evaluate vaccination status

Dietary/Herbal Supplements

- Are dietary or herbal supplements regulated?
 - A firm who manufactures or distributes is responsible for ensuring its safety
 - Dietary supplements do not need approval from FDA before they are marketed
 - FDA does not analyze dietary supplements before they are sold
 - FDA does make efforts to monitor the marketplace for products that are unsafe/false or misleading

How to Choose a Natural or Herbal Product?

- United States Pharmacopeia (USP) Verification Program
 - Voluntary program in which USP annually evaluates the quality of verified products
- The USP Verified Mark on the labels indicates that the product
 - Contains the ingredients listed on the label, in the declared potency and amount
 - Does not contain harmful ingredients
 - Has been made using safe and sanitary practices according to the FDA and USP guidelines



CF Nutrition: What I Learned from the Conference

- Important vitamins and minerals to help support health & well-being
- Nutrition recommendations for those on CFTR modulators
- Using body composition as part of the nutrition assessment

Vitamins and Minerals

Your body needs vitamins to help it grow, function, and fight off infection

- Consume foods rich in vitamins
- May need to take a vitamin supplement
 - Vitamin A,D,E,K (fat soluble vitamins)

Like vitamins, minerals also help with normal growth, function and maintenance of good health

- People with CF can be deficient in certain minerals
 - Calcium
 - Iron
 - Sodium chloride (salt) - for pediatrics
 - Zinc
 - May need supplementation if cannot get enough from diet

CF Nutrition and CFTR Modulators

- Since the introduction of CFTR modulators (Kalydeco®, Orkambi®, Symdeko®, and Trikafta®):
 - Many people with CF have experienced an increase in their weight
 - Weight gain can vary: minimal/moderate weight gain vs more significant weight gain.
 - Studies are being done to provide more specific information on how modulators affect nutrition and the digestive system

CF Nutrition and CFTR Modulators Continued

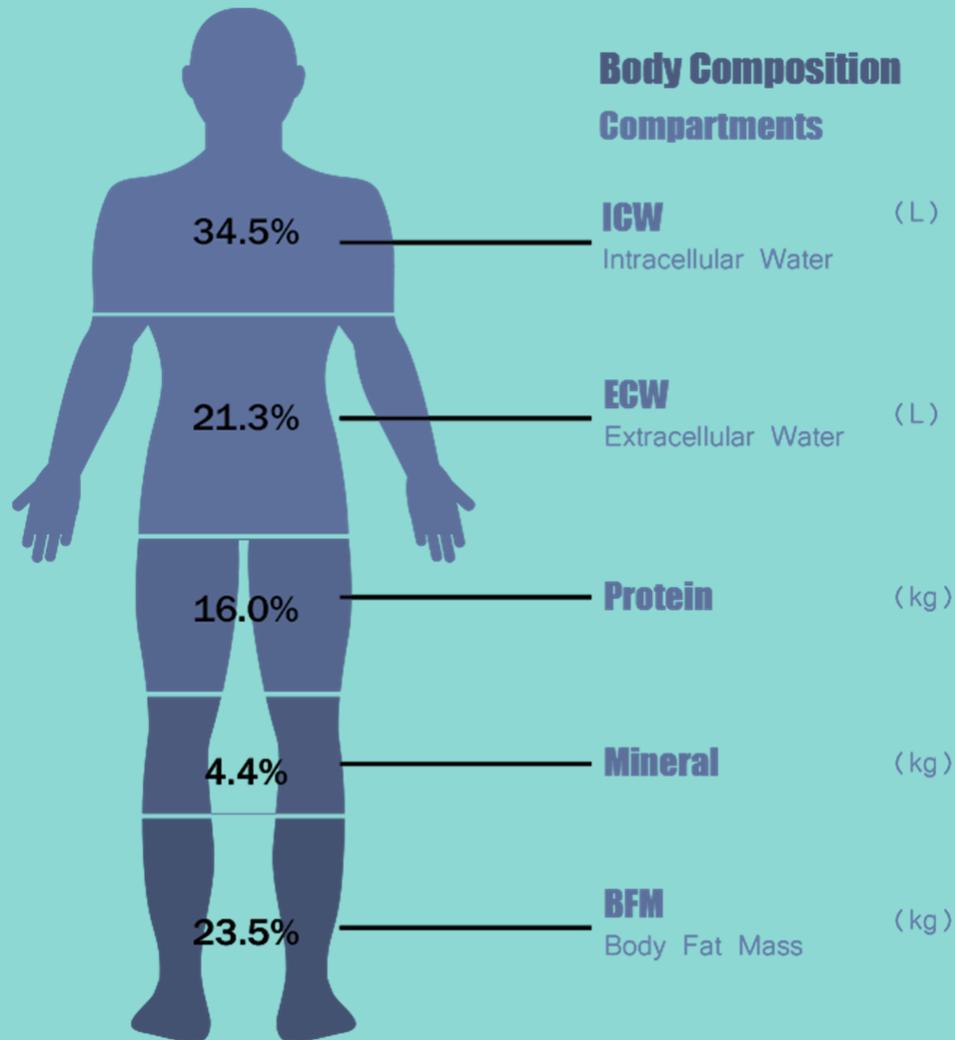
- Current recommendations:
 - Continue pancreatic enzymes (may need adjustments)
 - Eat foods that contain healthy unsaturated fats w/ modulators
 - i.e. string cheese, peanut butter, yogurt
 - Weight management Suggestions:
 - Decrease or stop your nutrition shake (Ensure, Boost, Pediasure, etc)
 - Decrease the number of snacks you eat daily
 - Replace some of your current foods with low calorie foods
 - Switch sugar-containing drinks to water
 - Move to low-fat or non-fat dairy products (except when using to take your Trikafta®)
 - Partake in physical activity as tolerated

Body Composition

- Goal for children with CF: Weight-for-length or BMI at/above the 50th percentile
- Goal for adults with CF: BMI of 22 kg/m² for females and BMI of 23 kg/m² for males
- Many CF patients have had weight gain with CFTR modulators
 - Focus on type of weight gain
 - Fat mass vs muscle mass
 - Skin fold test
 - Bioimpedance analysis
 - Underwater weighing
 - Air displacement (BodPod)



Body Composition Continued



Body Fat Percentage

Typical % for	Men	Women
Essential Fat	2-5%	10-13%
Athletes	6-13%	14-20%
Fitness	14-17%	21-24%
Average	18-24%	25-31%
Obese	25%+	32%+

Christina Bond, MSW, LMSW



My First NACFC

The biggest take away I had from NACFC 2022

It is such an exciting time to
work with our patients with

Cystic Fibrosis!



Tidbits from NACFC

Managing stress with chronic illness



Benefits of **Palliative Care** for those with CF

Bright Beginnings, Brilliant Futures is an online space to empower families with tools to live full lives and feel hopeful about their children's future.

Dialectical Behavioral Therapy - DBT used as an effective method of de-escalation during difficult interactions with patients

Acceptance & Commitment Therapy for Teens

TACTIC - Teens Acceptance and Commitment Therapy Involving CF.

This was a pilot program that consisted of a 6 week group, 60 minute session via Zoom with home assignments, weekly topic discussion, experiential activity and an on-theme mindfulness activity.

6 Core Processes of ACT discussed during 6-program

1. Acceptance
2. Cognitive Defusion
3. Contacting the Present Moment
4. Self as Context
5. Values
6. Committed Action

Dialectical Behavioral Therapy - DBT

This is an evidence based psychotherapy that began with efforts to treat personality disorders and interpersonal conflict;. It is a modified type of Cognitive Behavioral Therapy (CBT).

The main goals of DBT are to teach people how to live in the moment, develop healthy ways to cope with stress, regulate their emotions, and improve their relationships with others.

Disorders of emotional and behavioral regulation (borderline personality disorder, bipolar disorder, etc.) as well as self-destructive behaviors (eating disorders, substance abuse, self-harm, etc.) are commonly treated with DBT.

The main components are mindfulness and distress tolerance

Deb Fueller FNPC



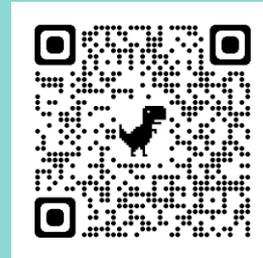
Share Endlessly, Steal Shamelessly

All information and slides are happily stolen from the amazing speakers presenting at the North American Cystic Fibrosis Conference.

To have the opportunity to choose your own topic please go to:

https://www.youtube.com/results?search_query=nacfc+2022

or scan the QR code:



The slides are a small representation of what I took from the conference. There is so much to take in and remember. So much excitement from the scientists as they reveal their breakthroughs.

And most importantly there is so much **HOPE** for the future.

There is sadness for those who were not able to experience the fruit of these labors. It brightens me to know that you can celebrate in the knowledge that participation in the CFF Registry study along with the brave individuals who directly participated in the treatment studies assisted in bringing these advancements forward .

5 main points

I focused most of my attention on the upcoming research and changes noted since the modulator therapy was started.

1. Significant decline in hospitalizations
2. increased life expectancy
3. significant increase in pregnancies
4. decreased number of lung transplants
5. Overall less problems with low weight (body mass index) and poor nutrition

Plenary Session One - Dr. Steven Rowe

(highly recommend viewing the Plenary sessions)

New Fronts with CFTR Modulator Therapy

The Current CF Modulator Therapy is credited for many things:

The effects of highly effective modulators:

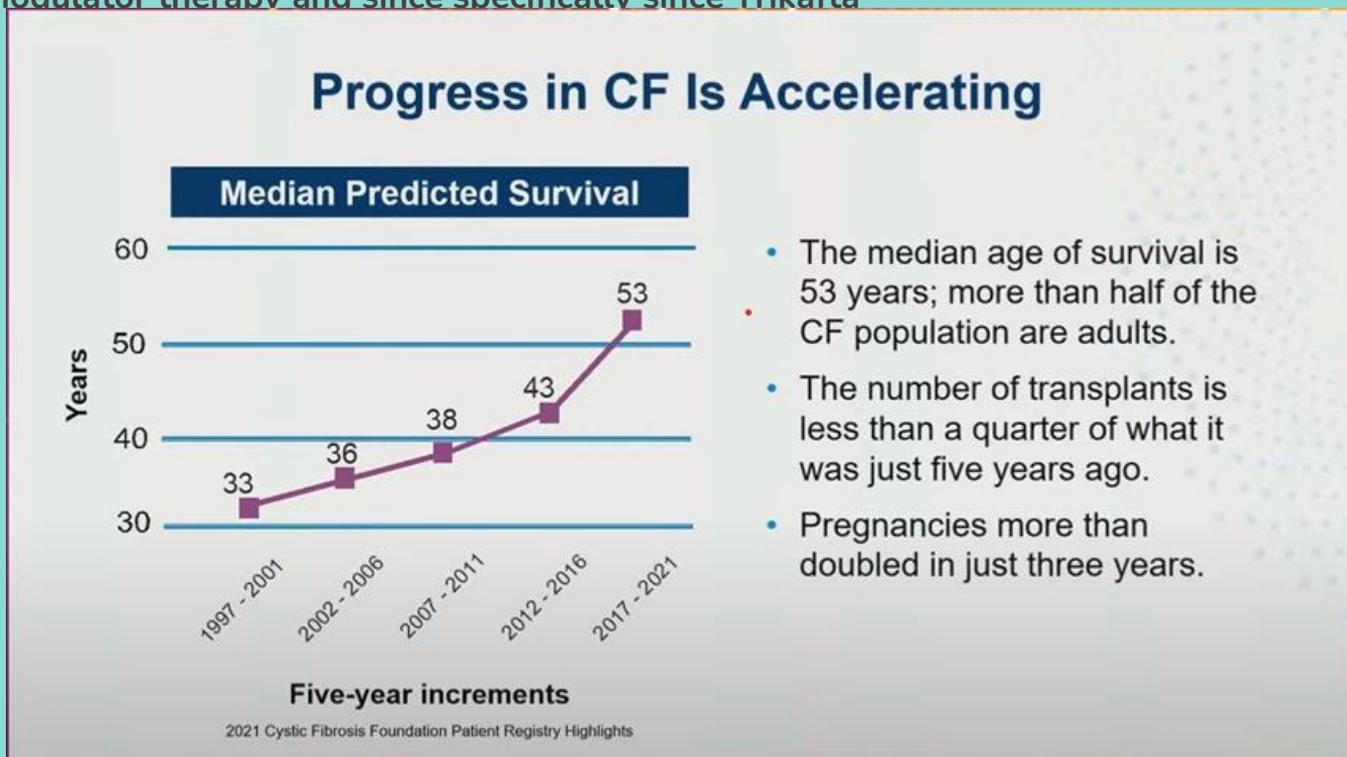
- Prolong survival
- Improve lung function
- Reduce pulmonary exacerbations
- Improve respiratory symptoms
- Reduce rate of lung function decline
- Augment mucus clearance
- Benefit sputum microbiology
- Reduce pulmonary inflammation
- Improve weight gain
- Accelerate linear growth
- Reduce sweat chloride
- Augment sweat rate
- Normalize GI pH
- Preserve pancreatic function

Currently the modulator therapy is able to treat about 90% of individuals with CF.

There is active research in seeking treatment for the other 10%

Plenary Session One

This session highlighted the predicted survival rate which is rising and showing a sharp upward trend with modulator therapy and since specifically since Trikafta



Treatment yes! No Cure Yet!

There is continued work on the path toward a cure with many different research studies ongoing.



Hope for All

Significant progress in individual health but the current therapy does not fit all variants.

Multiple trials going forward in an attempt to find treatment for those who are not currently in the treatment path of modulator therapy.

Checkout <https://apps.cff.org/trials/pipeline>

The number of drug studies to restore CFTR Function include:

PreClinical phase	12 trials	Phase 1
1 trial		
Phase 2	3 trials	
Phase 3	1 trial	

4 Modulators have completed trials and are out for patient use

Fertility

Every one of the US CF centers experienced a dramatic increase in pregnancies

Both in planned and unplanned pregnancies

Need for increased monitoring for those pregnancies.

Some anecdotal evidence of use of modulator therapies during pregnancy in nonCF mothers to treat issues of the CF positive baby.

Looking into possible changes in men with CF and potential for improved fertility.

Plenary Session 3 - Dr. Elizabeth Tullis

Evolving with the Times



How to Prepare for the Future as Individuals with CF Age

We need the help of those with CF:

What Can **People With CF** Do?



Communicate
goals and
preferences



Be open
to change



Partner
with CF team to
self-monitor



Participate
in clinical trials
and research



How to Prepare for the Future as Individuals with CF Age

The CF TEAM:

Reflect - Listen and recognise different needs. Update advice

Plan - For larger adult clinics. Fewer admissions.

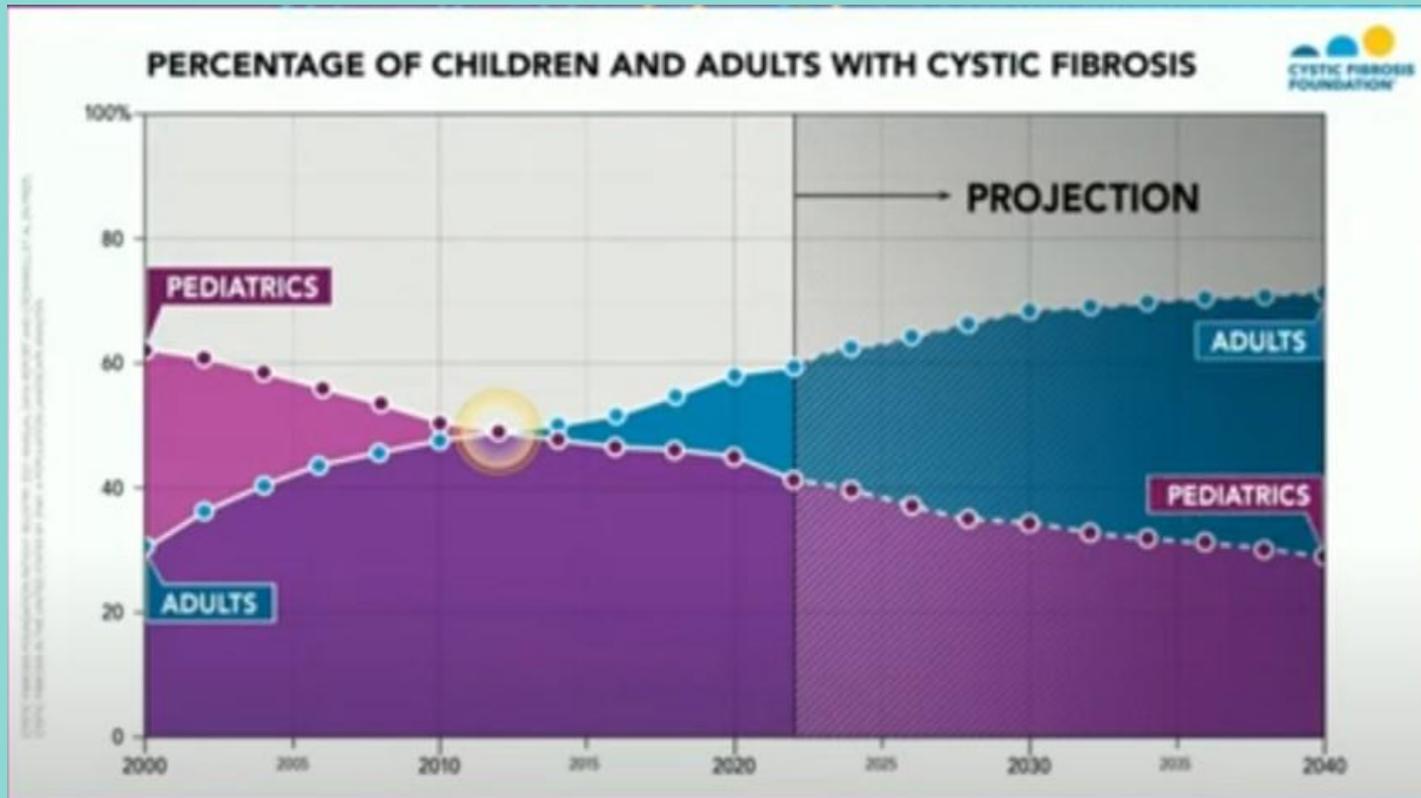
Readjust - Changes in the Team with specialties added.

More Social Workers, mental health providers and support staff.

Adapt - Shifting roles to accommodate the shifting needs

Inform - Center Directors to communicate with administration about upcoming changes.

Changing Makeup of Those with CF



Aging

Need for monitoring:

cholesterol levels

colonoscopies

prostate

blood pressure

mammography

Insurance changes as many enter the age of social security and face challenges of continuing medications not previously seen with frequency in medicare in the over 65 age group.

The Future of Cystic Fibrosis

Things to remember:

We continue to work toward a cure!! Watch the CFF Drug Pipeline to see the progress!

Treatment options have improved dramatically and the average age has risen

Need for ongoing therapy is being studied but no reduction in airway clearance therapies have been recommended

Lung damage that has occurred cannot be reversed

Stay in touch with your CF Team!! They are excited to see you to help when needed and celebrate every success!!

