SCHOOL NURSE Workshop

Cystic Fibrosis

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Children's of Alabama®

CYSTIC FIBROSIS

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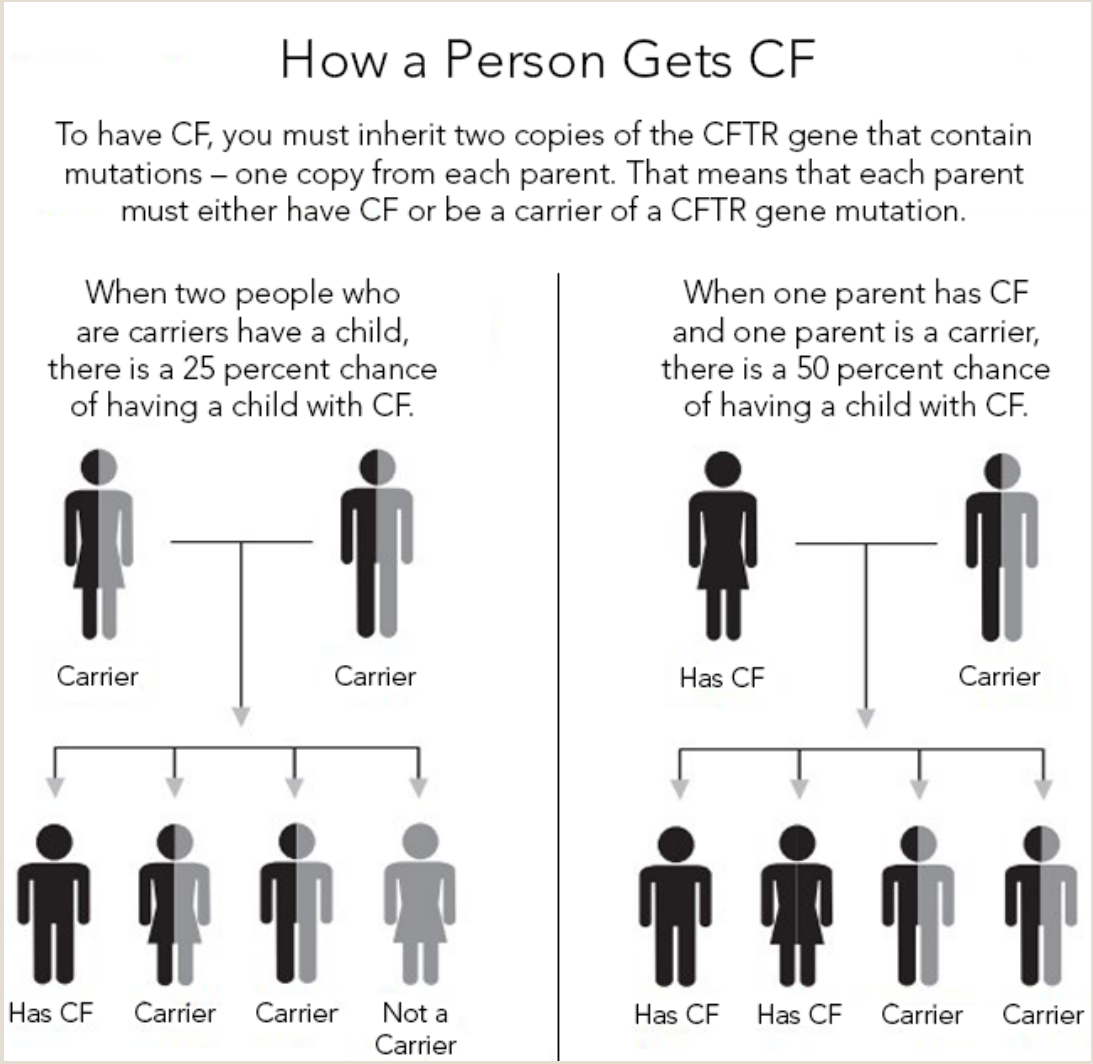
Objectives

- Understand the root cause of Cystic fibrosis (CF) and how it impacts the body
- Describe how CF is diagnosed
- Know the needs of a child with CF at school
- Become familiar with the available highly effective Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) modulator medications

CF is a recessive genetic disease



https://www.cff.org/intro-cf/cf-genetics-basics

Diagnosing CF

2 disease causing mutations with a positive sweat chloride test

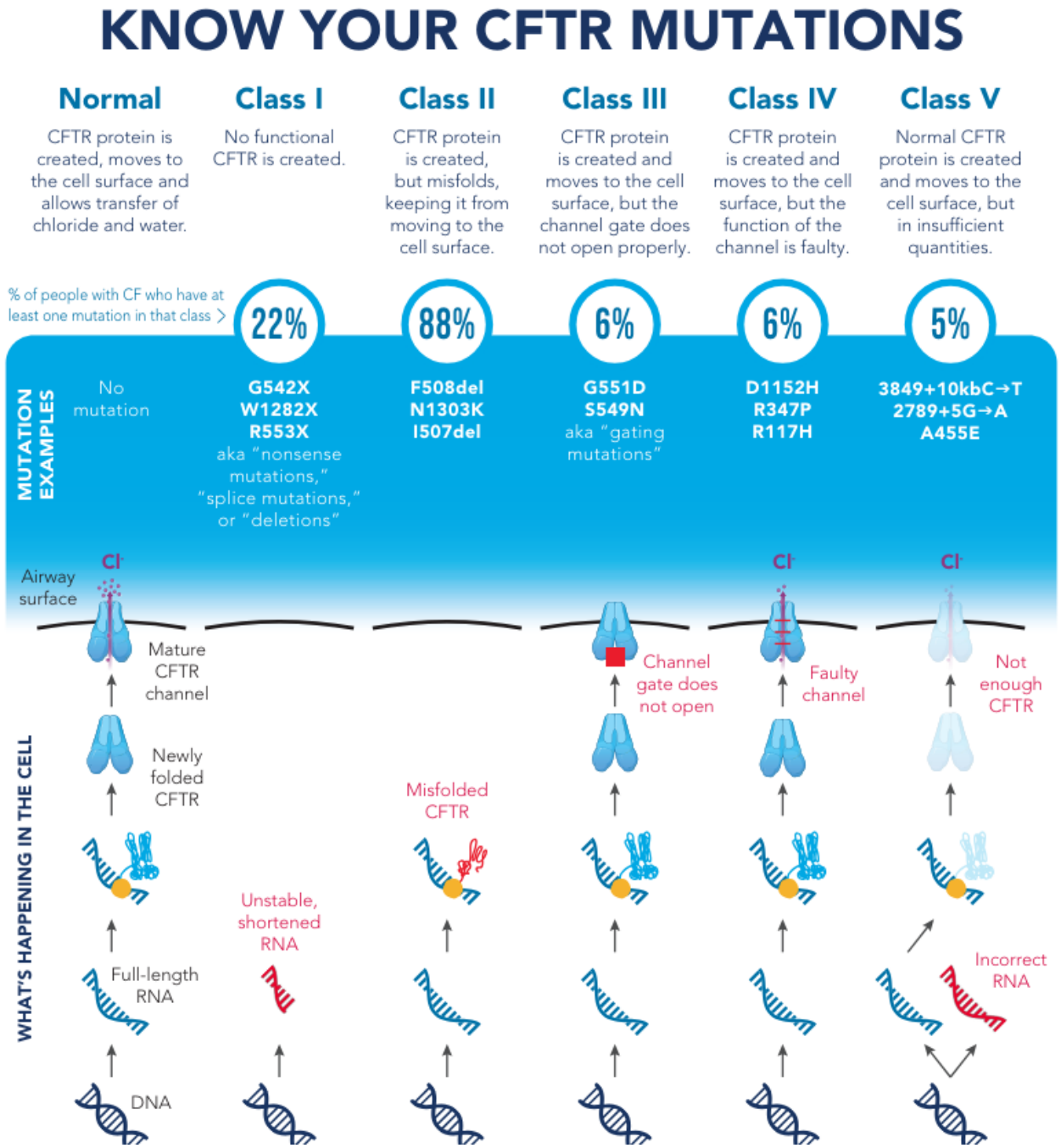
Genetic Variants

- Over 2000 CF causing variants have been discovered
- The most common CFTR variant is Delta F508

https://cftr2.org/resources

What is CFTR?

- CFTR = cystic fibrosis transmembrane conductance regulator
- Mutations in the CFTR gene cause the CFTR protein to malfunction or not be made at all



Newborn Screening (NBS)

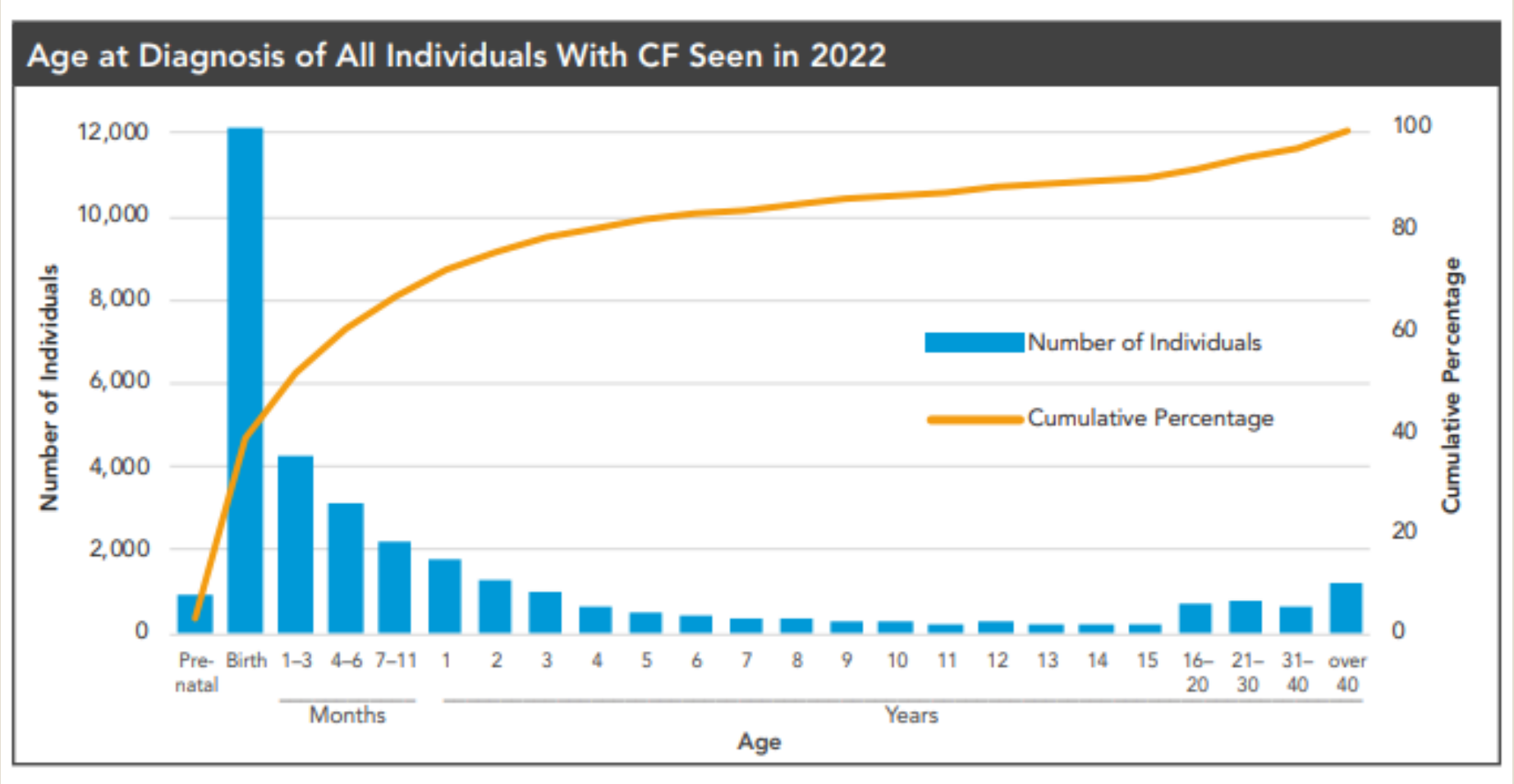
- Screened at birth – Immunoreactive Trypsinogen (IRT)/DNA
- Positive screens are referred to the Children's of Alabama CF NBS program with a sweat test
- Current Rate at Children's of Alabama: ~10-12 new diagnoses/year

CF population in the USA (2022)

- ~32,621 people in the US with CF
  - 752 of these patients were new diagnoses
    - 59.8% of these patients were diagnosed through newborn screening
    - Median age at diagnosis = 3 months

https://www.cff.org/managing-cf/understanding-changes-life-expectancy#how-long-are-people-with-cf-living-today

Age at diagnosis



https://www.cff.org/medical-professionals/patient-registry

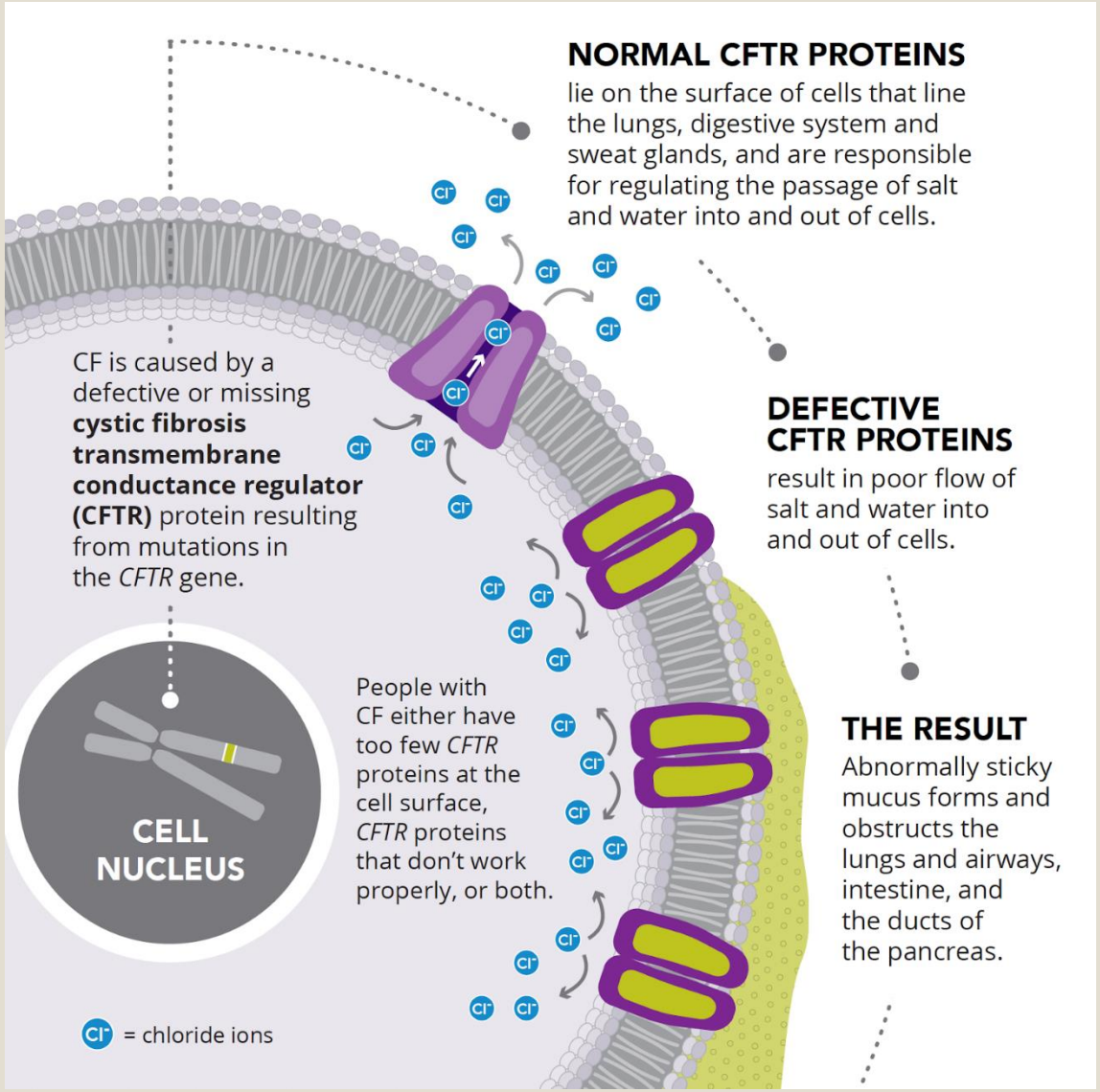
CF population in the USA (2022)

- 59.4% adult vs 40.6% pediatric
- Race/Ethnicity
  - 91.2% white
  - 3.5% African American
  - 5.3% other races
  - 10% Hispanic

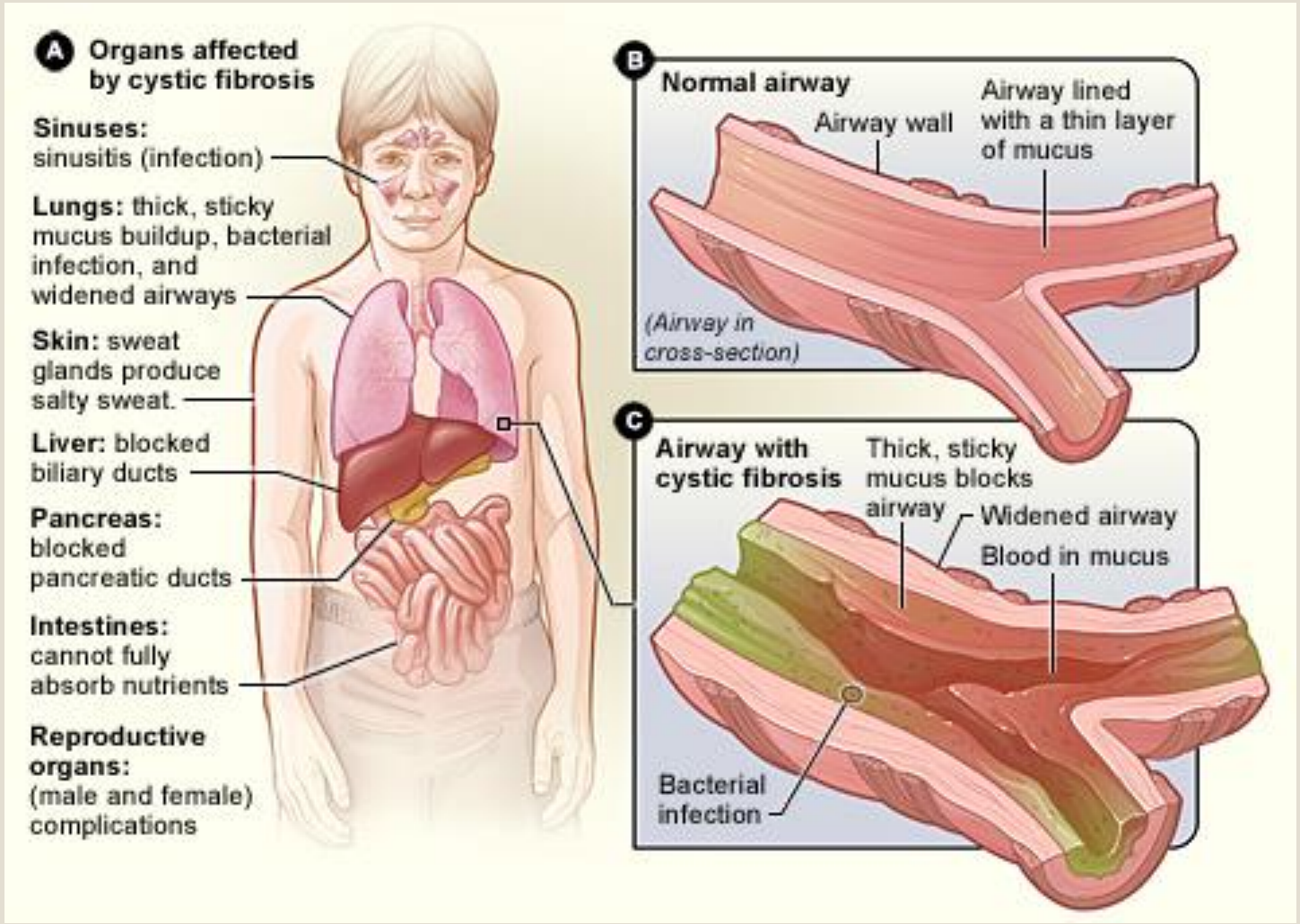
https://www.cff.org/managing-cf/understanding-changes-life-expectancy#how-long-are-people-with-cf-living-today

What Happens?

- CFTR regulates the proper flow of water and chloride into the cell
- When chloride does not flow through the gate, the balance of chloride and fluids is disrupted
- Mucus becomes thick and sticky in many organs

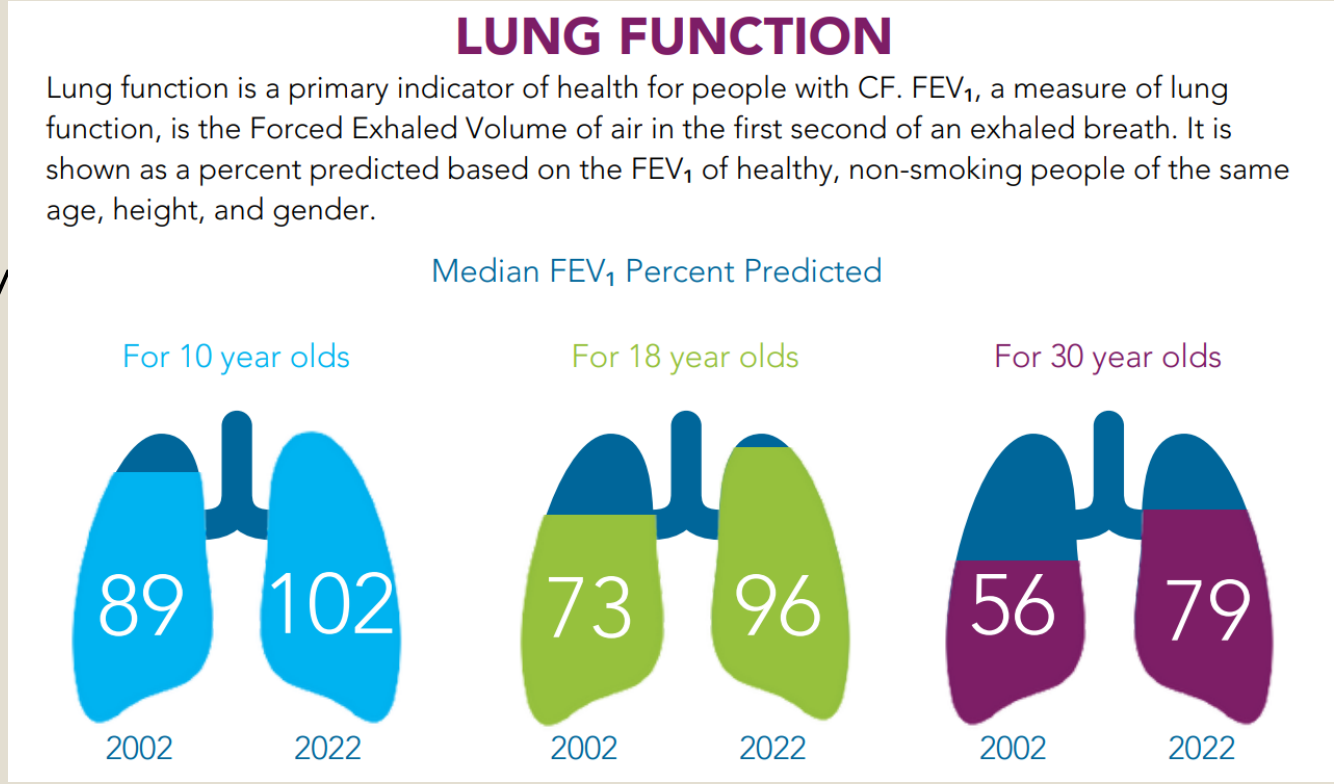


How CF Impacts the Body



Complications

- Reduced lung function
- Sinus Disease
- Dehydration
- Exocrine pancreatic insufficiency
- Difficulty gaining weight
- Cystic Fibrosis Related Diabetes
- Constipation/intestinal blockage
- Liver Disease
- Anxiety/depression



https://www.cff.org/sites/default/files/2023-05/2022-Cystic-Fibrosis-Foundation-Patient-Registry-Highlights-Handout.pdf

Highly Effective Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) Modulators

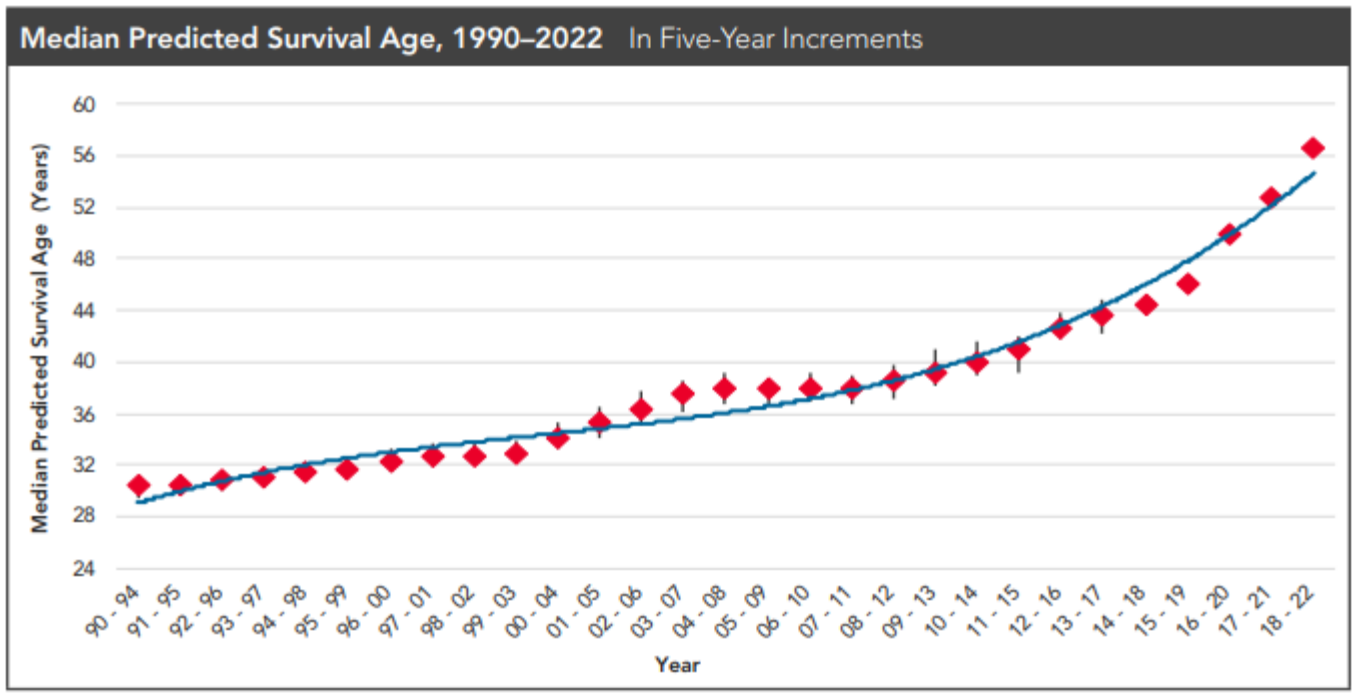
- Ivacaftor (Kalydeco®) - Age 1 month or older
- Elexacaftor/Tezacaftor/Ivacaftor (Trikafta®) - Age 2 years or older

General Positive Impacts of CFTR Modulators

- Improvement in lung function
- Decreased Sweat Chloride values
- Decreased cough/mucous
- Weight gain
- Improved quality of life
- Fewer pulmonary exacerbations requiring antibiotics

Median Predicted Survival

"For individuals born between 2018 and 2022, the median predicted survival age was 56.6 years."



*Using the currently recommended method for calculating median predicted survival. For more information about the methodology, please see the Technical Supplement available at cff.org.

https://www.cff.org/managing-cf/understanding-changes-life-expectancy#how-long-are-people-with-cf-living-today

CF Care (clinic)

- CF visit every 4-8 weeks for the first 2 years of life
- At best CF clinic visit every 3 months
- Annual labs: check vitamin levels, blood sugar, kidney and liver function
- Chest x-ray every 2 years
- Annual oral glucose test starting at 10 years old
- Pulmonary functions at every visit starting at 4 years old
- Other possible tests: DEXA (bone scan), Chest CT, Sinus CT, and Liver ultrasound

CF Daily Care (home)

- Chest therapy twice a day
- CFTR modulator medication twice a day
- Eat a balanced, healthy diet
- Take enzymes with every meal
- Take vitamins everyday
- Use other medications as prescribed (bronchodilators, mucolytics, inhaled or oral antibiotics, inhaled corticosteroids, etc.)
- Get plenty of rest and exercise

CF and school

- Medications
  - Pancreatic enzymes before meals and snacks
  - Bronchodilator as needed

CF and School

- Nutrition
  - May need extra portions of the entrée at meals
  - May need high calorie nutritional supplements
  - Allow access to water throughout the day
  - Pancreatic enzymes before meals and snacks

- Exercise is encouraged for people with CF.

https://www.cff.org/intro-cf/teachers-guide-cystic-fibrosis#:~:text=Accommodations%20may%20include%20allowing%20the,and%20meal%20to%20increase%20calories.

CF and School

- Infection Prevention and Control
  - Good hand hygiene for all
  - Cover mouth when coughing or sneezing for all
  - Encourage vaccinations
  - If there are 2 or more people in the same school with CF, minimize time spent in the same place and always maintain a 6+foot distance from one another.
  - Avoid allowing 2 people with CF to share common items including workstations, eating areas, bathrooms, etc. even if not using them at the same time.

https://www.cff.org/intro-cf/teachers-guide-cystic-fibrosis#:~:text=Accommodations%20may%20include%20allowing%20the,and%20meal%20to%20increase%20calories.

CF and School

- Other accommodations that may be needed:
  - Increased absences
  - Increased access/flexible bathroom breaks
  - Ability to leave the classroom during a coughing spell
  - IEP - Individualized Education Programs

https://www.cff.org/intro-cf/teachers-guide-cystic-fibrosis#:~:text=Accommodations%20may%20include%20allowing%20the,and%20meal%20to%20increase%20calories.

CF and School

- The school packet provided by the CF team at Children's of Alabama includes:
  - A teacher's guide to CF handout
  - School diet prescription
  - School letter summarizing the needs of children with CF
  - Medication forms (pancreatic enzymes and albuterol)

What can help children with CF to be successful in school?

- Educate all staff
- Practice good infection control
- 504 plan or IEP
- Ongoing communication between school staff and the student/family
- Involvement of the CF care team when needed

Blog post from a school nurse with 3 children with CF in her school

”The head of the guidance department and I decided to start by creating schedules for these three students…. Once their classes had been scheduled, I then needed to think about other circumstances such as lunchtime, hallways, extracurricular activities, assemblies, shared computer use and visits to the nurse's office. Next, I had a brief meeting with each student and their families for input. My ultimate goal is for all of my students to have a safe and memorable high school experience, and these three students are no exception.”

“Fortunately, my research and preparation appear to have paid off. One major reason things have gone so we[illegible]nazing staff and administration.”

Rachel Jackson, RN

https://www.cff.org/community-posts/2016-08/how-one-school-nurse-preparing-three-students-cf-same-school
https://www.cff.org/community-posts/2017-03/updates-school-nurse-three-students-cf-one-school

Education and Employment (2022)

EMPLOYMENT

57%

of adults with CF have full-time or part-time jobs.

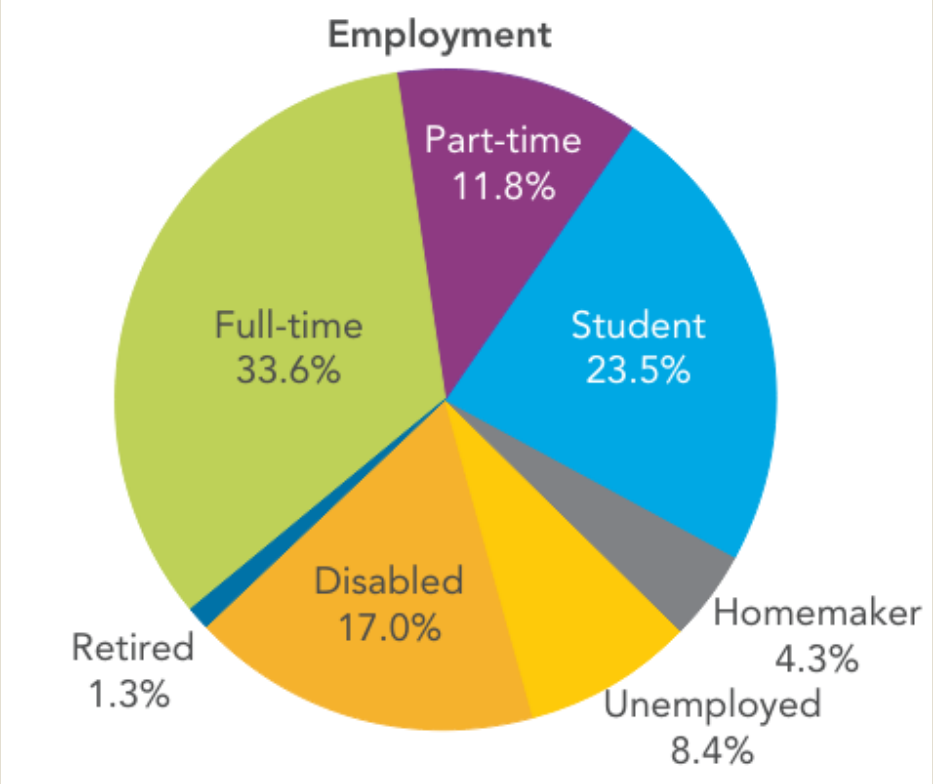
EDUCATION

41%

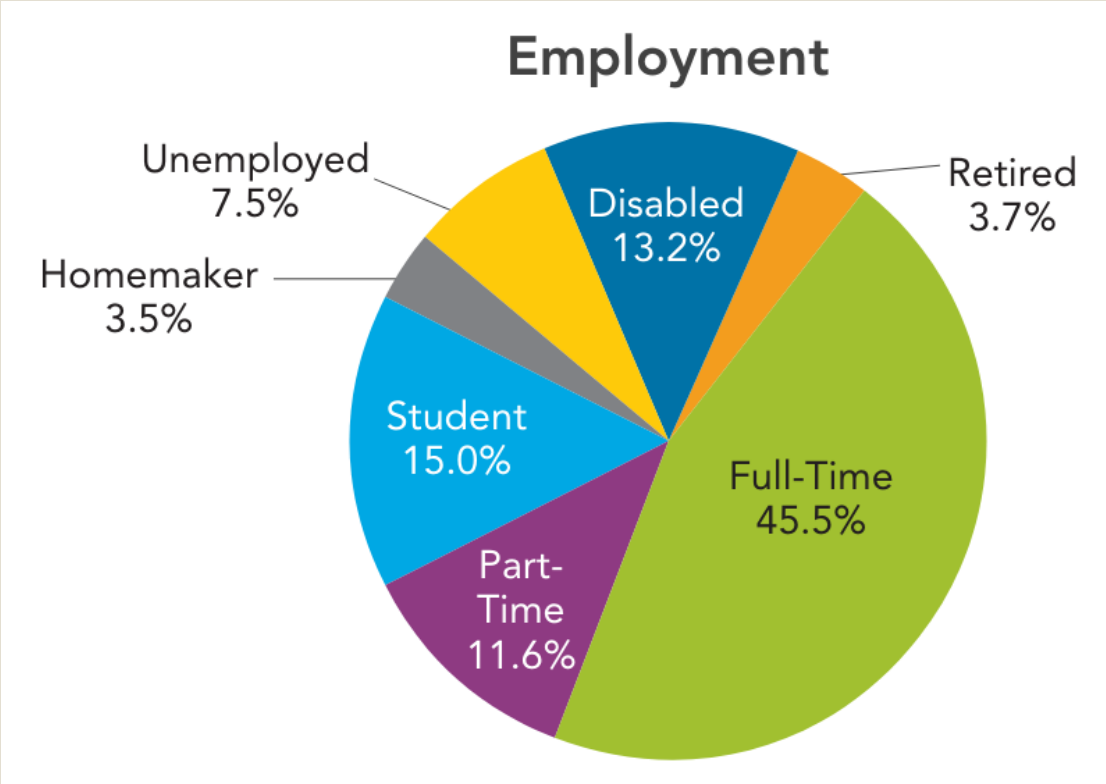
of adults with CF have a college degree.

2012→2022 Employment Rates for adults with CF

2012

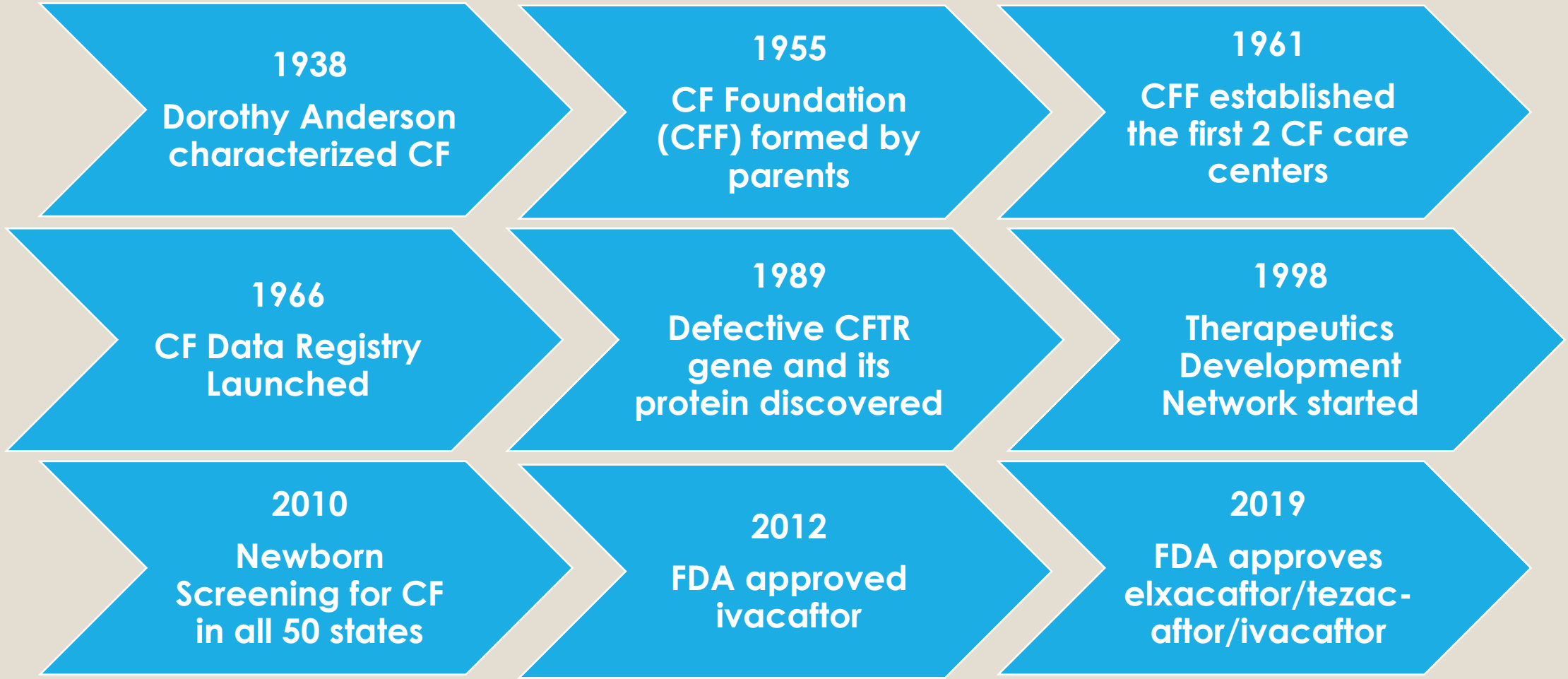


2022



11.7% increase in employment
3.8% decrease in disability
2.4% increase in retirement

History of Cystic Fibrosis (CF) - Milestones



https://www.cff.org/about-us/our-history

“Path to a Cure: Many Routes, One Mission”

- Repairing the CFTR Protein (CFTR Modulators) – patient will still have CF
- Restoring CFTR protein production – patient will still have CF
- Fixing or Replacing the CFTR Gene – addresses the root cause of the disease and the patient will no longer have CF
- The ultimate goal:

https://www.cff.org/research-clinical-trials/path-cure-many-routes-one-mission

Sunshine School

For information about the sunshine school services, visit the following Children's of Alabama web page:

https://www.childrensal.org/services/junior-league-birmingham-sunshine-school

Sunshine School Contact information:

Children's of Alabama
Department of Family Services
Sunshine School
Phone: **205-638-9651**
Email: sunshineschool@childrensal.org

University of Alabama at Birmingham/Children's of Alabama Pediatric CF Center

Contact Information:

Phone: 205-638-9583

Fax: 205-638-2457

Questions

References

- www.cff.org
- https://cftr2.org/resources
- https://www.cff.org/medical-professionals/patient-registry