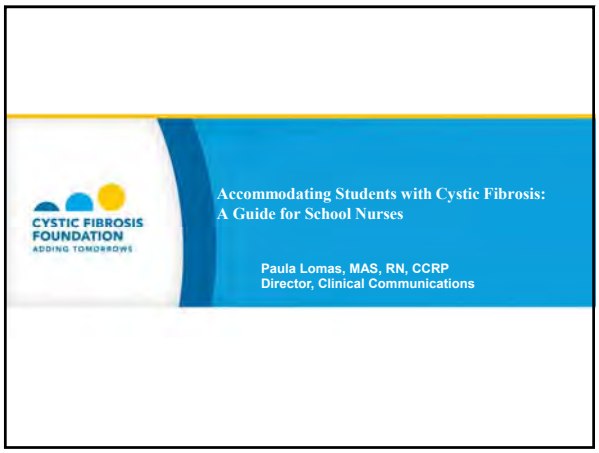


Objectives:

Attendees will be able to

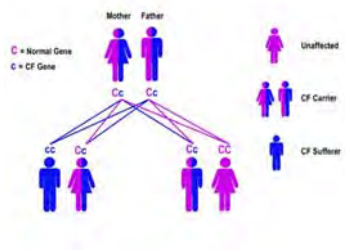
- Describe the diagnosis of cystic fibrosis and how it might affect the accommodations required for the school setting.
- Discuss the therapies required for a child with cystic fibrosis.
- Implement appropriate CF Foundation guidelines to your school setting.

Agenda

- What is cystic fibrosis?
- Treatments and therapies
- CF Foundation Guidelines

What is CF?

CF is the most common life-shortening autosomal recessive disorder in the US
- Affects about 33,000 people in the US and 70,000 worldwide
- 1 in 31 people are carriers



cff.org

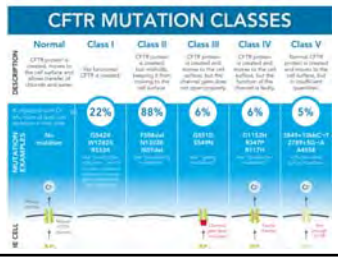
What is CF?

Now diagnosed primarily by newborn screen
- Some diagnoses are made in childhood and adulthood
- Confirmed through sweat test, genetic test and clinical evaluation

cff.org

What is CF?

Caused by mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene
- More than 2000 mutations (cftr2.org)
- Loss of CFTR function affects sodium and chloride ion channels, altering the flow of salt and fluids in and out of cells



What is CF?

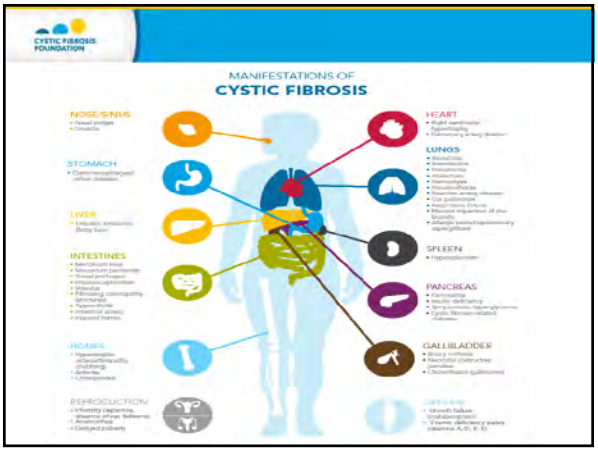
Thick, sticky mucus builds up and blocks the ducts of the exocrine glands, including the lungs and pancreas

- Mucus build up causes chronic infection, inflammation, and damage

Symptoms of CF

People with CF can have a variety of symptoms, including:

Very salty-tasting skin
Persistent coughing, at times with phlegm
Frequent lung infections including pneumonia or bronchitis
Wheezing or shortness of breath
Poor growth or weight gain in spite of a good appetite
Frequent greasy, bulky stools or difficulty with bowel movements
Male infertility



Case #1

During a conversation, a mother lets you know that her child is a carrier of the CF gene. You should:

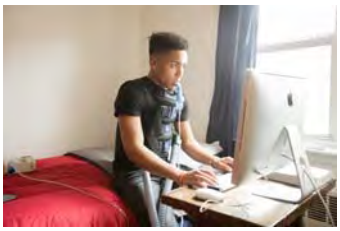
a) Make sure that you have a treatment plan for him.
b) Do nothing ✔
c) Learn more about CF and guidelines
d) Make sure you keep him away from other children with CF in your school

People with CF do best when:

- They have highly individualized treatment
- They can balance their treatments with daily life
- Their care plans address the whole person—physical, mental and social

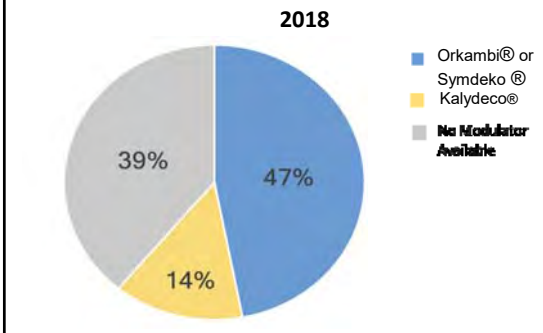
COORDINATING

Treatments and Therapies



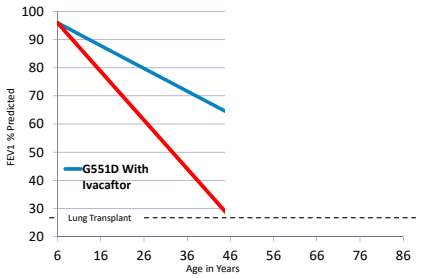
Highly effective CFTR Modulators Will Completely Change CF Care

CFTR Modulators



Rate of Lung Function Decline

Effect of decreased rate of decline in FEV_1



Next Generation CFTR Modulator Combinations Phase 2 Trial Results

- **Patients with <u>one copy of F508 del</u>**
  - **10%+ absolute improvement in FEV_1 compared to placebo**
  - 30+ mmol/L drop in sweat chloride
  - Remarkable improvement in quality of life
- **Patients with <u>two copies of F508del</u>**
  - **9% absolute improvement in FEV_1 compared to Tezacaftor and Ivacaftor**
  - 30+ mmol/L drop in sweat chloride
- **No safety concerns noted**

CYSTIC FIBROSIS FOUNDATION

CF is leading the medical field in "personalized" medicine

Work on "Rare Mutations"

- There are over 1000 CFTR mutations that 5 or less people in the world have
- We can't do standard clinical trials to make sure everyone who would benefit will receive drug

CYSTIC FIBROSIS FOUNDATION

Even with CFTR Modulators, a whole generation will have to deal with lung disease

In 10 years, over 50% of patients in the U.S. will still need symptomatic therapies despite potent new CFTR therapies

CYSTIC FIBROSIS FOUNDATION

You can maximize your student's overall learning experience, while helping to maintain his or her health.

Airway Clearance

To be performed 2-3 times per day

- Bronchodilator
- Mucus thinners
  - Hypertonic saline
  - Pulmozyme® (Dornase Alfa)
- Airway clearance technique
  - Manual chest physical therapy
  - High-frequency chest wall oscillation (vest)
  - Positive expiratory pressure (PEP) or oscillating PEP
  - Autogenic drainage
- Inhaled antibiotics
- Inhaled corticosteroids

Cough at school

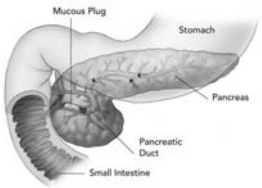
- Chronic
- Moves mucus out of large airway
- Not contagious
- Self conscious

In school accommodation:

- Nebulization treatment
- Airway clearance
  - Child can and should bring school work with them during the required treatment.
- Water, tissues, hand sanitizer at desk

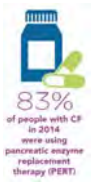
Nutrition

- Increase caloric requirement
- Obstruction of digestive system
  - Require vitamins
  - High-calorie, high-fat foods
- CF-related diabetes
- Gastroesophageal reflux
- Oral enzymes
  - Steatorrhea
  - Abdominal pain



Enzymes

- Aid in digestion of carbohydrates, protein and fats
- Brand necessary
- Individualized by dosage and brand
  - Lipase (digest fat)
  - Protease (digest protein)
  - Amylase (digest carbohydrate)
- To be given just before eating (meals and snacks)
- Room temperature



In school accommodation:

- Enzymes
- Caloric requirement of child with CF
- Bathroom usage
  - Freedom
  - Privacy
  - Odor

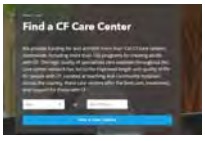
Pulmonary Exacerbation

- Chronic bacterial infections
- Acute exacerbation

Staphylococcus aureus (Staph)
Pseudomonas (P. aeruginosa)
Methicillin-Resistant *Staphylococcus aureus* (MRSA)
Burkholderia cepacia (B. cepacia)
Nontuberculous Mycobacteria (NTM)

In school accommodation:

- Partner with the child's care center

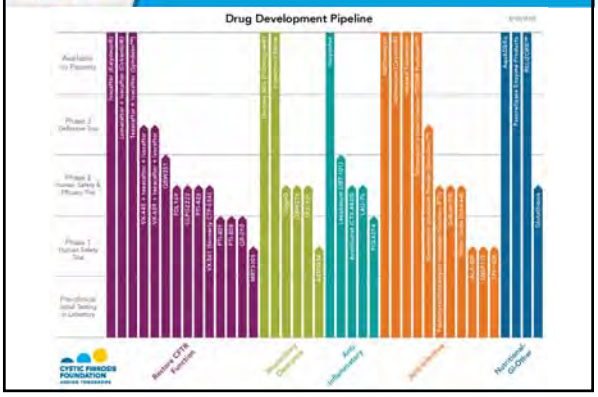


- Individualized Education Plans (IEPs)

IEP/504 plans

- Time needed for medication or therapy
- Adjust school rules to allow child to self administer enzymes
- Have a plan when child is hospitalized or home sick
- Audio or video tapes of class when missed
- Access to bathroom in nurses office
- Don't limit bathroom or water
- School medical emergency plan

Clinical Trials



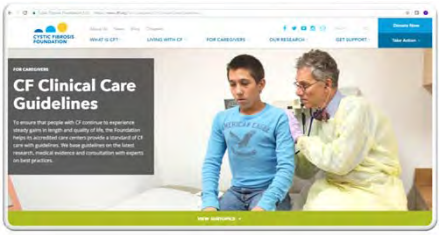
Case #2

Sam is in 5th grade and constantly forgets to come to the office for his enzymes. You should:

a) See if you can work out a new plan ✓
b) Do nothing
c) Give them to him after he eats lunch
d) Recommend that he take his enzymes before coming to school.

IMPROVING

Clinical Care Guidelines



www.cff.org

Infection Prevention and Control

Being Germ Smart

- Stay a safe 6 feet away from someone else who may be sick
- Hand hygiene: especially when using common items
- Avoid sharing personal items
- Cough hygiene

Infection Prevention and Control Guideline for Cystic Fibrosis: 2013 Update Saiman L. et al. *Infect Control Hosp Epidemiol.* 2014 Aug;35 Suppl 1:S1-S67. Epub 2014 Jul 1.PMID: 25025126

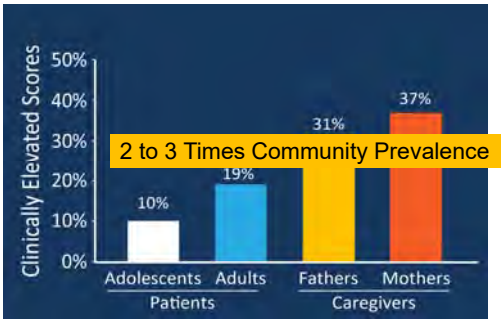
When there is more than one child with CF in your school

- Encourage disclosure
- Minimize the time in the same area (6 ft rule)
  - Separate classrooms
  - Separate bathrooms and water fountains
  - Schedule common areas (gym, cafeteria) at different times
  - Assign lunch tables, lockers as far away from each other as possible
  - Make sure not at nurses office at the same time
  - Make sure teachers know to notify nurse when sending student with CF to the nurses office

Resources@cff.org

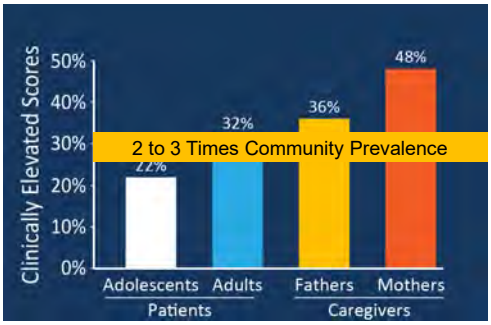
Mental Health

TIDES: Prevalence of Depression



Quittner et al. Thorax. 2014;69(12):1090-7.

TIDES: Prevalence of Anxiety



Quittner et al. Thorax. 2014;69(12):1090-7.

Impacts of Cystic Fibrosis

Mental Health (Depression)

- Clinic Attendance
- Exacerbations
- Lung Function
- BMI
- Quality of Life

CF related diabetes (CFRD)

- Not type I or type II
- Like type I
  - Insulin deficient
  - Due to scaring of pancreas
- Like type II
  - Insulin resistant
  - Could be due to inflammation , infection, steroid usage

Moran, A. et. al. Diabetes Care 2010. Dec, 33(12):2697-2708

CF related diabetes (CFRD)

- Not treated with diet and exercise
- Not treated with orals
- Do not limit calories to 1200!
- May have times when don't need insulin

Moran, A. et. al. Diabetes Care 2010. Dec, 33(12):2697-2708

Symptoms CFRD

- Polyuria
- Polydipsia
- Lethargy
- **Weight loss**
- Ketoacidosis is not usually associated with CFRD

Annual screening: OGTT beginning at age 10

Case #3

You have 2 children in your school with cystic fibrosis.
You should:

a) Screen them for anxiety and depression
b) Review documents on cff.org
c) Make sure their lockers are far apart
d) B and C ✓
e) All of the above

BELONGING

CYSTIC FIBROSIS FOUNDATION

Mission

To cure CF and provide all people with the disease the opportunity to lead full, productive lives by funding research and drug development, promoting individualized treatment and ensuring access to high-quality, specialized care.

https://www.cff.org/About-Us/About-the-Cystic-Fibrosis-Foundation/Our-Mission/

Accredited Care Centers

- Comprised of pediatric and adult care programs and affiliate programs
- 126 centers in the United States
- 285 Programs:
  - 123 pediatric
  - 110 adult
  - 52 affiliates

https://www.cff.org/Care-Centers/Find-a-CF-Care-Center/

Treatment Burden

Therapy	Time per treatment	Frequency (minimum)	Total time spent daily
Inhaled bronchodilators (such as β-adrenergic agonists, anticholinergics)	5 minutes	2–3 times/day	10–15 minutes
Inhaled mucolytics such as • hypertonic saline • dornase alfa (Pulmozyme)	20 minutes 5 minutes	2 times/day 1 time/day	40 minutes 5 minutes
Inhaled antibiotics such as • tobramycin (TOBI), colistimethate (Coly-Mycin M) • aztreonam (Cayston)	20 minutes 3 minutes	2 times/day 3 times/day	40 minutes 9 minutes
Airway clearance[a]	20–30 minutes	2 times/day	40–60 minutes

Lomas, P. Fowler, S., AJN, August 2010 - Volume 110 - Issue 8 - p 30-37

Partnerships for Sustaining Daily Care

Questions? Call 1.800.FIGHT.CF

Case #4

You now have a better understanding of challenges of cystic fibrosis. When you have a child with CF in your school you should:

a) Encourage the parents to connect you with their care team.
b) Review documents on cff.org
c) Feel free to call 1.800.FIGHT.CF with questions
d) All of the above ✓

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