*the Child Dental Patient with*

# Cystic Fibrosis (CF)

# CF- Epidemiology

- > 70,000 individuals worldwide;
- >1000/ year for Caucasians; most common terminal genetic disorder among Caucasians
- ~1 in 2,500 among Northern Europeans (the highest prevalence);
- disproportionately affects Southern Europeans, Ashkenazi Jews, African Americans.

# CF - Etiology

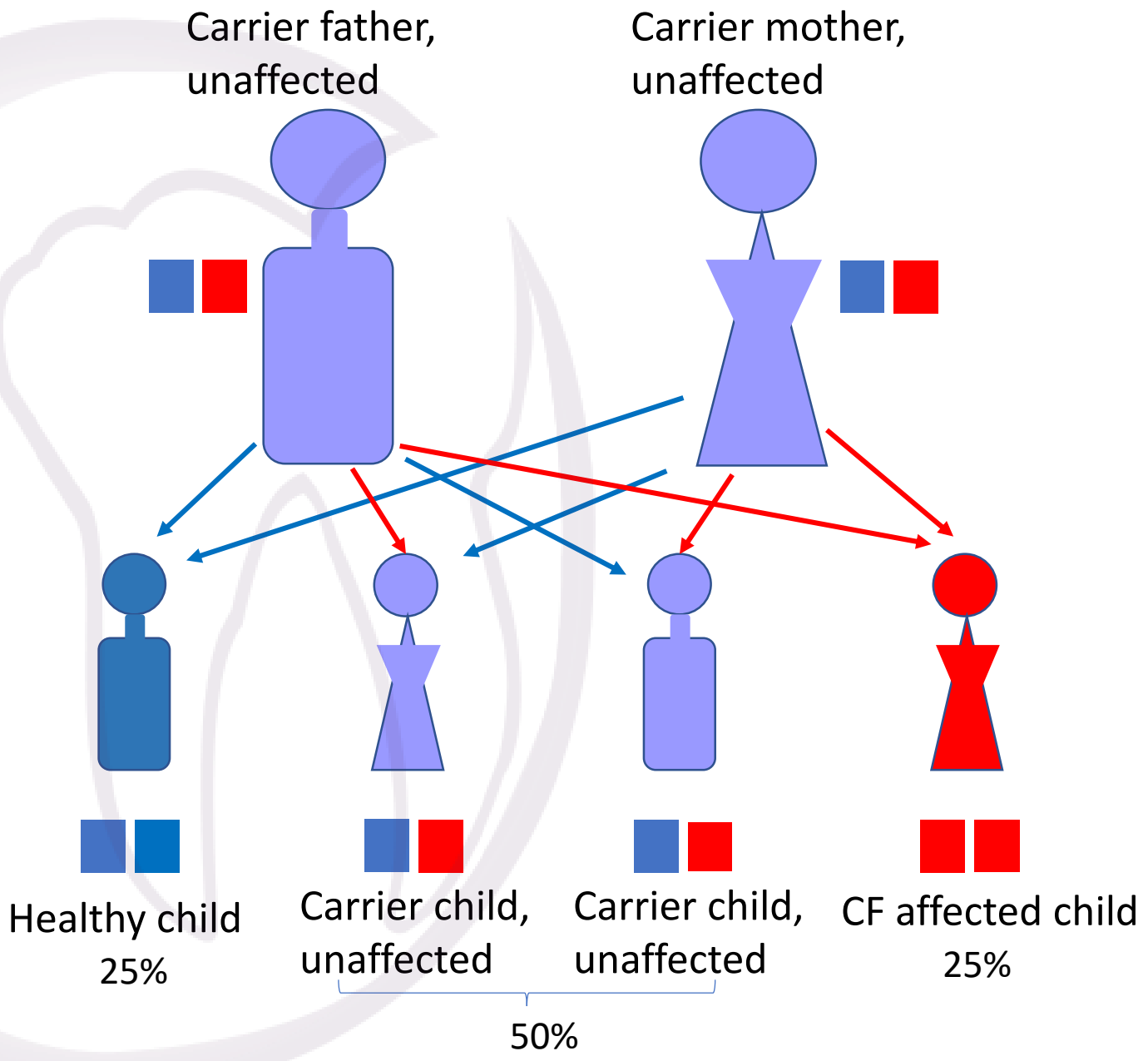
Multiple mutations of a single gene (**Cystic Fibrosis Transmembrane Regulator [CFTR])** on chromosome 7

→affects regulation of cellular chloride channels.

# Autosomal recessive transmission of CF:

*If each parent has a normal CFTR gene and a mutated CFTR gene (unaffected carriers), each child will have (regardless the gender):*

- *25 % chance of inheriting two normal genes*
- *50 % chance of inheriting one normal CFTR gene and one mutated CFTR gene and being a CF carrier*
- *25 % chance of inheriting two genes with mutations and having CF.*



# CF - Symptoms

- 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.

# CF - Characteristics

- **chronic airway inflammation**
- **mucus plugging**
- **bronchial infection** with specific bacterial pathogens

# Cystic Fibrosis

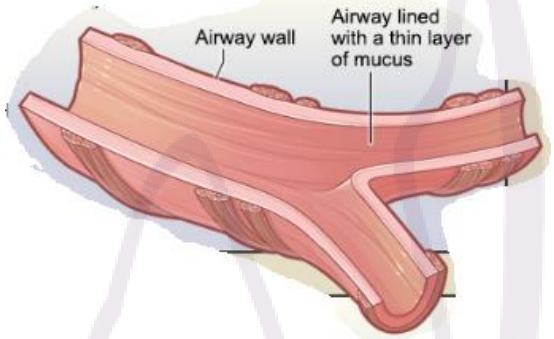
## Oral signs

- Disorder of salivary glands
- Swelling lips
- Gingivitis
- Dryness

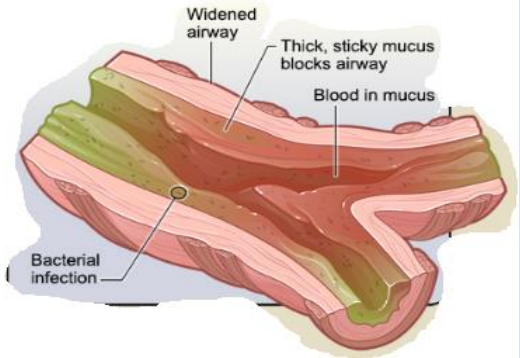
## Organs affected by cystic fibrosis

- Sinuses: sinusitis (infection)
- Lungs: thick, sticky mucus buildup, bacterial infection and widened airways
- Skin: sweat glands produce salty sweat
- Liver: blocked biliary ducts
- Pancreas: blocked pancreatic ducts
- Intestines: cannot fully absorb nutrients
- Reproductive organs: problems with fertility or delayed puberty

## Normal airway




## Airway with cystic fibrosis




**SOURCE:** National Heart, Lung, and Blood Institute; National Institutes of Health; U.S. Department of Health and Human Services (NIH): https://www.nhlbi.nih.gov/health-topics/cystic-fibrosis

# CF therapy - Pharmacological management

Treatment of **bronchial inflammation** and **infection:**

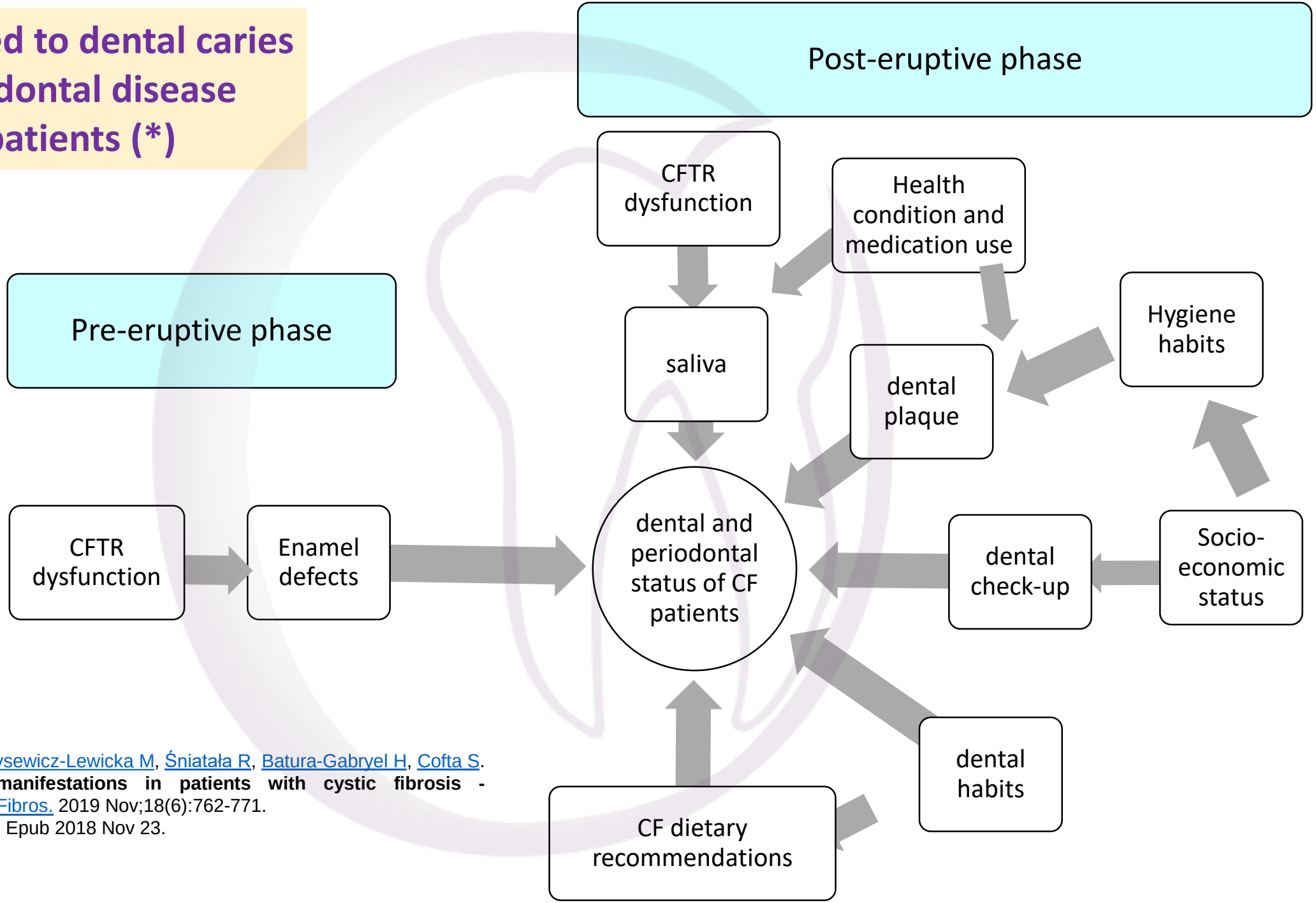
- **antibiotics** and **physiotherapy;**
- **pancreatic enzyme** replacement therapy;
- **fat-soluble vitamin supplementation;**
- $\beta_2$ **agonists** and **corticosteroids.**

# CF - Dietary Recommendations

to maintain the increased calories intake needed:

- **high calorie diet;**
- **high sugary foods.**

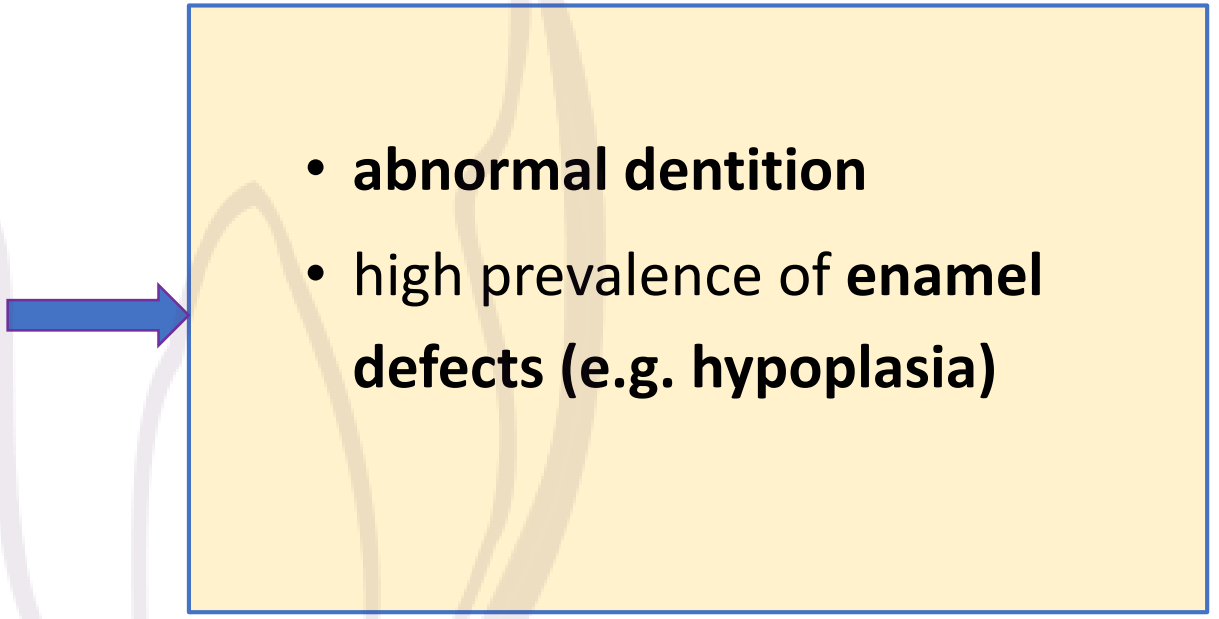
# Factors related to dental caries and periodontal disease in CF patients (*)




(*) Source:
Pawlaczyk-Kamieńska T, Borysewicz-Lewicka M, Śniatała R, Batura-Gabryel H, Cofta S. **Dental and periodontal manifestations in patients with cystic fibrosis - A systematic review.** J Cyst Fibros. 2019 Nov;18(6):762-771. doi: 10.1016/j.jcf.2018.11.007. Epub 2018 Nov 23.

# Oral health related-problems

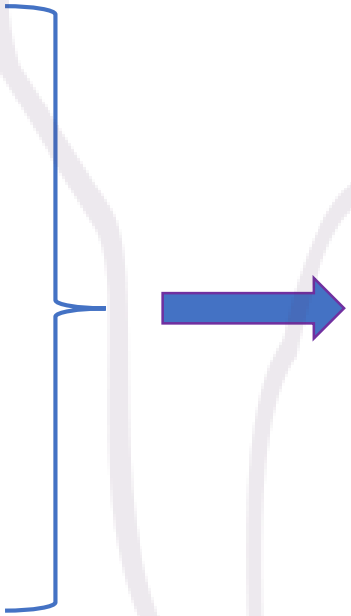
- CFTR gene- enamel formation role
- metabolic disease
- +++long-term pharmacological therapies




**CFTR gene** is highly expressed in salivary glands, but the reported effects of CF on **salivary gland function** and the incidence of **dental caries** are inconsistent.

# Cystic Fibrosis – Oral Findings

- **Higher pH**
- saliva **buffering capacity**



- **Low caries** prevalence

## Oral management of children with CF

- multidisciplinary approach team
- oral health should be under control since early years of life by paediatric dentists

- The American Dental Association (ADA) and the American Academy of Paediatric Dentistry (AAPD) recommend **dental check-ups every 6 months,** with more frequent dental check-ups for children at increased risk for caries.

During dental check-ups:

•assess the patient's **oral health**
•**monitor growth and development**
•evaluate the need for any treatment
•make **recommendations to reduce caries risk**

→

+++ *Preventive care*:
•**dental cleanings**
•**oral hygiene instructions**
•**dietary counselling**
•**pit-and-fissure sealants**
•**topical fluoride applications**

# Important for the Paediatric Dentist

- The paedodontist should **consult with the patient's physician** prior to rendering treatment.
- Patients should be **treated in** a more **upright position** to allow them to clear secretions more easily.
- **Avoid** the use of **sedative agents** that interfere with pulmonary function.

## Important for the Paediatric Dentist

- Routine dental treatment rises few problems and there is **no contraindication to local anesthesia**.
- **Extractions** can be carried out **under local anesthesia** normally.
- For patients with severe pancreatic involvement, coagulation time of the blood should be checked before surgery, in view of the possibility of the lack of vitamin K.

## Important for the Paediatric Dentist

- **Discoloration of the teeth by tetracycline** is a serious problem to be faced in the dental care of these children.

  Most of them need **aesthetic improvement** as appearance is quite unacceptable.

  **Acrylic caps** appear as a good interim option for aesthetics improvement until the patient is old enough for full jacket crowns.

# Important for the Paediatric Dentist

- Children with cystic fibrosis have dry airways, and administration of **inhalation sedation can be dangerous** when the gases are not humidified.
- Moderate to severe pulmonary disease can be aggravated and degenerate to severe respiratory distress when inhalation anaesthetics are used.
- **General anesthesia (GA)** is essentially a matter for **in-patient care (hospital setting)**, and should **never be attempted in the dental surgery** except where the pulmonary involvement is negligible and the patient's physician has been consulted.
- GA is also problematic because concurrent administration of anticholinergic drugs further aggravates airway dryness. If GA is required for dental care, the patient should be hospitalized and managed by an anesthesiologist.

# Conclusion

- Cystic Fibrosis is not very common in children but whenever pediatric dentists come across the case, they should be able to manage them with precautions to improve the quality of life of these children.

- Before advances in antibiotic therapy, physical therapy and nutritional supplementation are important to increase the median life expectancy of children with CF.