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# Determining the Topical Curative and Subsequent Access of Cystic Fibrosis

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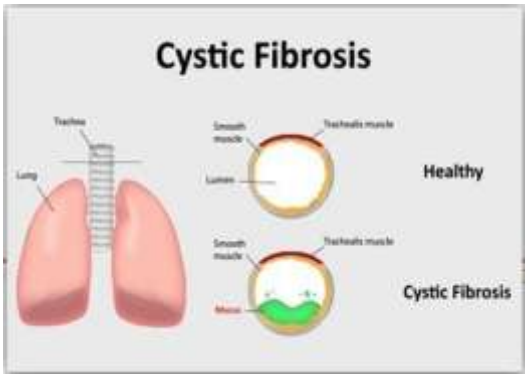
**ABSTRACT:**
Cystic fibrosis is the most prevalent inherited disease caused by a defect in the cystic fibrosis transmembrane conductance regulator (CFTR) gene. Hopes of preventing this cascade ofevents are provided by the development of new therapies that address the underlying defect of airway dehydration.
CFTR dysfunction affects many organs; however, lung disease is responsible for the vast majority of morbidity and mortality in patients with cystic fibrosis. Prenatal diagnostics, newborn screening and new treatment algorithms are changing the incidence and the prevalence of the disease.

**Keywords:** Cystic fibrosis, CFTR, sweat, pancreatic insufficiency, pancreatic sufficiency, Chloride, childrens.

## I. INTRODUCTION:

Cystic fibrosis (CF) is an inherited disorder that causes severe damage to the lungs, digestive system and other organs in the body. Cystic fibrosis affects the cells that produce mucus, sweat and digestive juices.




The classical CF phenotype is quite complex, involving multiple epithelial lined organs, although lung manifestations are the dominant source of morbidity and mortality.

It is a chronic disease that frequently leads to chronic sinopulmonary infections and pancreatic insufficiency. CFTR protein is expressed in many cells and has several functions, not all of which have been linked with disease. The primary function of the CFTR protein is as an ion channel that regulates liquid volume on epithelial surfaces through chloride secretion and inhibition of sodium absorption.

**AIMS AND OBJECTIVES:**

To identify the effect of comparative with present approaches and future approachesin cystic fibrosis.
Study of currently approved drugs and exploration of future clinical development pipeline the rapeutics for cystic fibrosis and possible limitations in their use.

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**MECHANISM OF ACTION:**

- When the protein is not working correctly, chloride — a component of salt — becomes trapped in cells.
- Without the proper movement of chloride, water cannot hydrate the cellular surface.
- This leads the mucus covering the cells to become thick and sticky, causing many of thesymptoms associated with cystic fibrosis

**SIGNS AND SYMPTOMS:**

- persistent cough that produces thick mucus (sputum)
- Wheezing
- Exercise intolerance
- Repeated lung infections
- Inflamed nasal passages or a stuffy nose
- Recurrent sinusitis
- Frequent chest infection
- Shortness of breath
- Poor weight gain and growth
- Intestinal blockage, particularly in newborns

**Figure 1 signs and symptoms of cystic fibrosis**

**DIAGNOSIS:**

In one screening test, a blood sample is checked for higher than normal levels of a chemical called immunoreactive trypsinogen (IRT), which is released by the pancreas. A newborn's IRT levels may be high because of premature birth or a stressful delivery. For that reason, other tests may be needed to confirm a diagnosis of cystic fibrosis. To evaluate if an infant has cystic fibrosis, doctors may also conduct a sweat test once the infant is at least 2 weeks old. A sweat-producing chemical is applied to a small area of skin. Then the sweat is collected to test it and see if it's saltier than normal. Testing done at a care center accredited by the Cystic Fibrosis Foundation helps ensure reliable results. Doctors may also recommend genetic tests for specific defects on the gene responsible for cystic fibrosis. Genetic tests may be used in addition to checking the IRT levels to confirm the diagnosis.

**Immunoreactive trypsinogen (IRT) test.**

The IRT test is a standard newborn screening test that checks the blood for abnormal levels of the protein called IRT. A high level of IRT may be a sign of CF. However, further testing is required to confirm the diagnosis.

**Sweat chloride test.**

The sweat chloride test is the most commonly used test for diagnosing CF. It checks for increased levels of salt in the sweat. The test is performed by using a chemical that makes the skin sweat when triggered by a weak electric current. Sweat is collected on a pad or paper and then analyzed. A diagnosis of CF is made if the sweat is saltier than normal.

**TREATMENT:**

Possible treatments include:

- antibiotics to prevent and treat chest infections
- medicines to make the mucus in the lungs thinner and easier to cough up
- medicines to widen the airways and reduce inflammation
- special techniques and devices to help clear mucus from the lungs
- medicines that help the person absorb food better
- following a special diet and taking supplements to prevent malnutrition

**MEDICATION:**

- Co-trimoxazole.
- Tobramycin.
- Cephalexin.
- Colistin.
- Dicloxacillin.
- Azithromycin.
- Amoxicillin.

**PREVENTION:**

**PRIMARY PREVENTION:**

Vaccinations, counseling to change high-risk behaviors, andsometimes chemoprevention are types of primary prevention.

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**SECONDARY PREVENTION:**

In secondary prevention, disease is detected and treated early, often before symptoms are present, thereby minimizing serious consequences.

**TERITIARY PREVENTION:**

Tertiary prevention involves the prevention of complications in people who have already developed disease, and in whom disease prevention is no longer an option. For these patients, the goal of tertiary prevention is to maximize the outcomes and prevent further morbidity from the disease process.

## II. METHODOLOGY:

**Past view of cystic fibrosis:**

The past 20 years have transformed the field of CF treatment sothat changing the course of this fatal illness is becoming a reality. The view of the road ahead hasnever been so encouraging in CF therapeutics, and there is cause for unprecedented optimism foranyone who cares for people with this devastating disease.

Over past 5-6 decades with better understanding of pathogenesis of CF, significant improvement has occurred in treatment manifesting as survival benefit. The Cystic Fibrosis Foundation (CFF) has projected a life expectancy of 37 years for CF patients currently and a UK model predicts that a child born with CF today can expect to live past 50 years of age . In countries with limited resources like India, the survival of children with CF is improving but lags considerably behind the developed countries.

**Present view of cystic fibrosis:**

Today, there are **close to 40,000 people in the United States** with CFdue in large part to more people living longer into adulthood, signaling a shift in what it means tolive with CF in the modern era.

**There's currently no cure for cystic fibrosis**, but a number of treatments are available to help control the symptoms, prevent complications, and make the condition easier to live with. Possible treatments include: antibiotics to prevent and treat chest infections.

**Future view of cystic fibrosis:**

In the next decade, it is likely that both a CFTR potentiator (Kalydeco) and at least one CFTR corrector may be available for the majority of patients with CF.

Newer antimicrobial approaches will focus on disarming these pathogens before they can establish structures associated with persistence, such as biofilms. Because future therapies will aim to prevent, rather than improve, existing organ damage, one future challenge will be developing better measures of early organ dysfunction well before patientsbecome symptomatic.

## III. CONCLUSION:

Future therapies will aim to prevent, rather than improve, existing organ damage, one future challenge will be developing better measures of early organ dysfunctionwell before patients become symptomatic. Moreover, most of the molecules discussed above are still in early developmental or early clinical phase, with limited safety and efficacy data, and a lasting beneficial outcome cannot be guaranteed with any.

Currently available treatments are still not able to fully address the complexity of the disease. New treatments are urgently needed for cystic fibrosis patients who do not benefit from CFTR repair drugs. In addition, patients on CFTR drugs are still experiencing long-term lung function decline – this needs to be addressed via alternative drug mechanisms" which was needed for future treatment.

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