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The evolution of cystic fibrosis: therapeutic approaches for adult patients.

The evolution of cystic fibrosis: therapeutic approaches for adult patients

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Summary

Introduction: Cystic Fibrosis (CF), or mucoviscosidosis, is a hereditary disease with an autosomal recessive pattern and early onset, affecting children in their first years of life. Patients with this disease present with thick, viscous mucous secretions that obstruct the ducts of exocrine glands, contributing to three basic characteristics: chronic obstructive pulmonary disease, elevated sweat electrolyte levels, pancreatic insufficiency with maldigestion and malabsorption, and consequent secondary malnutrition. CF has seen an improvement in its prognosis in recent decades; thus, patients with the disease reach adulthood with well-established care for the pathology. This progress has been achieved through the implementation of multidisciplinary care in specialized centers, including the use of systemic and inhaled antibiotics, airway clearance therapy, pancreatic enzyme replacement, and a high-calorie diet. **General Objective:** The primary objective of this research is to discuss the pathology and the importance of specialized treatment. **Methodology:** As a methodology, a bibliographic survey will be used through the main online databases (PubMed, SciELO, BVS), using a filter to select the most relevant articles from the years 2006 to 2025. **Results:** It is noted that the development of CFTR (Cystic Fibrosis Transmembrane Conductance Regulator) protein modulators has improved pulmonary function and significantly increased the life expectancy of patients. **Conclusion:** It is concluded that early diagnosis, combined with an individualized therapeutic approach, is essential to improve the prognosis and provide a better quality of life for patients with Cystic Fibrosis.

Keywords: Cystic Fibrosis. Disease. Hereditary. Adults. Treatment.

Abstract

Introduction: Cystic Fibrosis (CF), also known as mucoviscidosis, is a hereditary disease with an autosomal recessive pattern and an early onset, affecting children in the first years of life. Patients with this condition produce thick and viscous mucus secretions, leading to obstruction of the ducts of exocrine glands. This contributes to three main characteristics: chronic obstructive lung disease, elevated electrolyte levels in sweat, and pancreatic insufficiency, resulting in poor digestion, malabsorption, and consequent secondary malnutrition. The prognosis of CF has improved over the past decades, allowing patients to reach adulthood with well-established care for the disease. This progress has been achieved through the implementation of multidisciplinary care in specialized centers, including the use of systemic and inhaled antibiotics, airway clearance therapy, pancreatic enzyme replacement, and a high-calorie diet. **Main objective:** The primary objective of the research is to discourse about the pathology and the importance of specialized treatment. **Materials and methods:** as methodology will be used the bibliographic survey through the main online collections (PubMed, SciElo, VHL), using a filter to select articles with greater relevance within from 2006 to 2025. **Results:** It is noted that the development of CFTR (Cystic Fibrosis Transmembrane Conductance Regulator) protein modulators improved lung function and significantly increased the life expectancy of patients. **Conclusion:** It is concluded that early diagnosis, combined with an individualized therapeutic approach, is essential to improve the prognosis and provide a better quality of life for patients with Cystic Fibrosis.

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1. Introduction

The thoracic cavity, oval in shape in cross-sectional diameter, is divided into three compartments: the right and left pulmonary cavities, which house the lungs and pleura, and the central mediastinum, which constitutes the third section and contains most of the other thoracic structures (GRAY, 2010). The lungs are enveloped by the pleura, which is divided into visceral pleura, when it covers the upper lungs. The entire pulmonary surface, and the parietal surface when it lines the pulmonary cavities. Among them, we find... if the pleural cavity, a space that contains pleural fluid, responsible for lubricating the surfaces and allow smooth sliding between the pleura. In addition, the lungs play They play an essential role in respiration and have an apex, a base, and three borders (inferior, posterior, and posterior). The right lung is subdivided into three lobes — anterior) and two surfaces (medial and costal).

Superior, middle, and inferior — separated by an oblique fissure and a horizontal fissure. The lung, on the other hand, is divided into three parts: superior, middle, and inferior — separated by an oblique fissure and a horizontal fissure.

The left brain is divided into two lobes, superior and inferior, by a single oblique fissure. (MOORE, 2019). In addition, the alveolar surface of the lungs is coated with surfactant, a substance which reduces surface tension, which is generally high (MOORE, 2019). In addition, some organs Organs involved in gastrointestinal function can be highlighted, such as the pancreas, the intestine, and the airways. The bile ducts and the liver. The pancreas, one of the largest glands in the digestive system, performs both functions. Both exocrine and endocrine glands secrete various enzymes responsible for lipid digestion.

Carbohydrates and proteins. The intestine is divided into the small intestine (duodenum, jejunum, and ileum) and the large intestine (cecum, (vermiform appendix, ascending, transverse, descending and sigmoid colons, rectum and anal canal), is the The main site for absorption of ingested nutrients, in addition to reabsorbing water from non-recyclable waste products. digested liquid chyme. The bile ducts, in turn, carry the bile produced in the liver to the The bile enters the duodenum and is stored in the gallbladder. Bile emulsifies fats, facilitating their absorption. Absorption occurs in the distal part of the intestine. Finally, the liver receives all the nutrients absorbed by the body. digestive system, with the exception of fats. (GRAY, 2010). In light of the above, it is observed that The organs mentioned are of fundamental importance in maintaining the body's homeostasis. ensuring proper functioning according to physiology. However, certain pathologies They can cause an imbalance in homeostasis, and can even be fatal if left undiagnosed. early onset. One example is Cystic Fibrosis (CF), a lethal genetic inherited disease. autosomal recessive, considered the second most common and with an incidence of Approximately 1 in every 2,500 live births, and in most cases, it is identified while the baby is still in the womb. childhood. (SCHNEIDERS, 2024). Cystic Fibrosis (CF) is caused by a mutation in the cystic gene. fibrosis transmembrane conductance regulator (CFTR), responsible for encoding a protein that It regulates the transmembrane conductance of chloride. This disease mainly affects the lungs and pancreas. and the gastrointestinal tract. The gene mutation alters the viscosity of secretions, resulting in poor nutrient absorption, electrolyte loss through sweat, pancreatic insufficiency, and changes in

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pulmonary secretions, due to the atypical transport of chloride and sodium through this protein. In

Over the last 70 years, Cystic Fibrosis has been recognized as the most significant hereditary disease.

which increased the need for early diagnosis. The disease can be detected through

The newborn screening test is performed, but confirmation is done through genetic testing. Because it is a disease that...

Early detection is required, and treatment is initiated immediately after diagnosis, aiming to improve the condition.

The prognosis improves, allowing patients to reach adulthood with specialized care.

The progress was made possible thanks to the implementation of multidisciplinary measures, such as the use of

Systemic and inhaled antibiotics, airway clearance therapy, enzyme replacement.

pancreatic diseases and a high-calorie diet. Therefore, to ensure a longer life expectancy, the

Proper treatment is essential for adults with the disease, highlighting the need for a

multidisciplinary approach.

2 theoretical frameworks

2.1 General objective

To conduct a literature review on the clinical aspects and main treatments of Cystic Fibrosis.

2.2 Specific objectives

To provide guidance on the importance of treating cystic fibrosis in adults, in order to improve...

improving their quality of life and increasing their life expectancy, highlighting the need for

A multidisciplinary approach to control symptoms, prevent infections, and treat complications.

Addressing the importance of adherence to treatment helps to slow the progression of the disease and its effects.

complications, such as respiratory failure, ensuring a longer and healthier life for them.

patients.

2.3 Theoretical Framework

Cystic Fibrosis (CF) is a genetic disease resulting from a mutation in

transmembrane conductance regulatory protein (CFTR), a channel expressed in the apical portion of

epithelial cells of various organs (ATHANAZIO et al., 2017). The disease occurs when the individual

inherits two variants of a particular gene, one from the father and one from the mother. The most common variant is

called the F508del variant. The CFTR gene controls the production of a protein that regulates the

movement of chloride, bicarbonate, and sodium (salt) across cell membranes; thus, with its

In this variation, the protein becomes dysfunctional, and if it doesn't function correctly, the movement of

Chloride, bicarbonate, and sodium is interrupted (ROSENSTEIN, 2021).

Cystic Fibrosis (CF) is a congenital, autosomal recessive disease, also known as

such as cystic fibrosis. The discovery of the CFTR gene in 1989 opened new possibilities for study.

about this condition (MATOS; MARTINS, 2020). The disease causes certain glands

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They produce abnormally thick secretions, 30 to 60 times more viscous than normal.
(BRAZIL. Ministry of Health. Cystic fibrosis. Brasília: Ministry of Health, 2024. Available at:
(https://bvsms.saude.gov.br/fibrose-cistica/. Accessed on: January 10, 2026).

Cystic fibrosis (CF) is a multisystemic disease that affects the pancreatic, hepatic, and other digestive systems.
Gastrointestinal and reproductive disorders are the primary cause of death, with impaired reproductive functions being the most common cause of death.
pulmonary (DUCATI, et al., 2024). In addition, defective alkalinization and dilution of the
Bile, obstruction of the bile ducts, inflammation of the biliary tree, and biliary fibrosis, which can progress
for multilobular fibrosis (ENAUD, et al., 2018). Among the most common symptoms are:
the inability to reabsorb chlorine, which results in its concentration in sweat (GABTZ; RITTER,
2007), malabsorption of nutrients, especially proteins and lipids, in addition to complications
Gastrointestinal conditions such as rectal prolapse, intestinal obstruction syndrome, constipation, and liver cirrhosis.
(ROSA, et al., 2008). Other symptoms include recurrent pneumonia and diarrhea (SCHNEIDERS,
2024).

It is estimated that around 80,000 people worldwide live with Cystic Fibrosis.
In North America, at least 31,411 people are being monitored for this condition, while in Brazil, the diagnosis is
confirmed in 5,773 individuals (OSWALDO CRUZ FOUNDATION (Fiocruz)).
Key questions about cystic fibrosis: how to diagnose it? Rio de Janeiro: Fernandes Institute
Figueira, 2023. Available at: https://portaldeboaspraticas.iff.fiocruz.br/atencao-crianca/principais-
questions-about-cystic-fibrosis-how-to-diagnose/. Accessed on: January 10, 2026).

Cystic fibrosis can be detected through the newborn screening test and diagnosed with...
Genetic testing. The test should be performed on samples collected within 30 days of the newborn's life.
born. To confirm suspected cases, the "Sweat Test" is used, which measures the amount of
Chlorides in sweat (SCHNEIDERS, 2024). All patients should undergo testing.
genetic studies, both to establish the necessary care and better guidance, and to clarify the genetic mutation
involved (OSWALDO CRUZ FOUNDATION (Fiocruz). Main
Questions about cystic fibrosis: how to diagnose it? Rio de Janeiro: Instituto Fernandes Figueira, 2023.
Available at: https://portaldeboaspraticas.iff.fiocruz.br/atencao-crianca/principais-questoes-sobre-
cystic fibrosis - how to diagnose/. Accessed on: January 10, 2026).

In addition to Cystic Fibrosis, the Newborn Screening Test also detects five other health problems.
Health: phenylketonuria, congenital hypothyroidism, sickle cell disease and other hemoglobinopathies, congenital
adrenal hyperplasia, and biotinidase deficiency (FRASÃO, 2024). Currently, Brazil
It has a comprehensive neonatal screening program for Cystic Fibrosis, in addition to...
Reference centers spread across most states for patient follow-up.
(ATHANAZIO, et al., 2017).

Because it is a complex and unique disease, CF requires a specific approach in its treatment.

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Treatment involves a multidisciplinary team working in centers with specialized therapies (MATOS; MARTINS, 2020). Conducting investigations is of utmost importance.
additional ways to improve these therapies, which may be relevant to other diseases (AMARAL, 2016).

Thus, interventions include antibiotics, bronchodilators, and medications to thin the bloodstream. pulmonary secretions, treatments for clearing airways in case of problems respiratory, pancreatic enzyme supplements and vitamins for digestive problems, as well as medications to improve cystic fibrosis protein function in people with certain conditions genetic variants (ROSENSTEIN, 2021). Lung transplants are indicated in cases of chronic obstructive pulmonary disease, primary or secondary pulmonary fibrosis, bronchiectasis, Pulmonary hypertension and in very severe cases of cystic fibrosis, regardless of etiology (FRASÃO, 2021).

The disease currently has no cure, but if diagnosed early, it can be treated. significantly reducing its effects on the body. Physical exercise should be done part of the treatment, according to the patient's capabilities. They contribute to the improvement of Respiratory function, help with muscle mass gain, control diabetes related to fibrosis. They help prevent cystic fibrosis and osteoporosis, as well as correct some acquired postural problems. due to respiratory problems (SCHNEIDERS, 2024)

3. Materials and methods

The main objective of this literature review was the analysis and collection of data and information previously published in scientific articles, books and dissertations, originating from The main sources of information that address Cystic Fibrosis were used. For this purpose, collections were utilized. Online databases such as PubMed, SciELO, and the Virtual Health Library (VHL) were analyzed. Published literature was also reviewed. in the main health information sources, namely: SciELO, PubMed and Virtual Library. Health (BVS), in addition to government websites. The references went through a selection filter, those chosen were those published between 2006 and 2025 that are most relevant. scientific.

4. Results and discussion

The results demonstrate that the development of CFTR protein modulators (Cystic Fibrosis Transmembrane Conductance Regulator) is associated with a significant improvement. of pulmonary function in patients with cystic fibrosis. A consistent increase was observed in Respiratory function parameters, accompanied by a reduction in the frequency of exacerbations. pulmonary problems and the need for hospitalization. Additionally, therapy with modulators of

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CFTR contributed to the improvement of the overall clinical status and quality of life of patients. These

Positive clinical effects are reflected in a significant increase in life expectancy.

highlighting the impact of CFTR modulators on disease progression.

Final considerations

The findings of this study indicate that early diagnosis of cystic fibrosis, associated with

An individualized therapeutic approach plays a key role in improving

Clinical prognosis of patients. Early identification of the disease enables the implementation of...

timely targeted interventions, including the use of CFTR protein modulators, therapies

respiratory support and nutritional strategies contribute to the preservation of lung function.

and to reduce complications throughout the course of the disease. Furthermore, the personalization of

Treatment, based on genetic profile and individual clinical manifestations, has proven to be

crucial for optimizing therapeutic outcomes, resulting in significant gains in

Quality of life and patient survival. These results reinforce the importance of policies.

neonatal screening, continuous multidisciplinary follow-up, and expanded access to therapies

Innovative approaches to the management of cystic fibrosis.

REFERENCES

ALVES, Stella Pegoraro; FRANK, Márcia de Azevedo; BUENO, Denise. Medications used in a pediatric population with cystic fibrosis. Einstein (São Paulo), v. 16, n. 4, eAO4212, 2018.

ATHANAZIO, RA; SILVA, LVRF DA; VERGARA, AA; RIBEIRO, AF; RIEDI, CA; PROCIANOY, EFA; ADDE, FV; REIS, FJC; RIBEIRO, JD; TORRES, LA; FUCCIO, MB; EPIFANIO, M.; FIRMIDA, MC; DAMACENO, N.; LUDWIG NETO, N.; MARÓSTICA, PJC; RACHED, SZ; MELO, SFO *Brazilian guidelines for the diagnosis and treatment of cystic fibrosis.* Brazilian Journal of Pulmonology, v. 43, no. 3, p. 219–245, 2017.

BURGEL, PR *What do adults with cystic fibrosis want from their doctors?* Chest, vol. 162, no. 6, p. 1225–1226, 2022.

COSTAGUTA, G.; PATEY, N.; ÁLVAREZ, F. *Cystic fibrosis liver disease in children: a review of our current understanding.* Archivos Argentinos de Pediatría, v. 121, no. 4, e202202905, 2023.

DE BOECK, K.; AMARAL, MD *Progress in therapies for cystic fibrosis.* The Lancet Respiratory Medicine, vol. 4, no. 8, p. 662–674, 2016.

DUCATI, GC; CARDOSO, J.; FERRAZEANE, EP; SCHIVINSKI, CIS *Respiratory system parameters in children with low severity cystic fibrosis: is there early involvement in relation to healthy peers?* Revista Paulista de Pediatria, v. 42, e2023030, 2024.

ENAUD, R.; FRISON, E.; MISSONNIER, S. et al. *Cystic fibrosis and noninvasive liver fibrosis assessment methods in children.* Pediatric Research, vol. 91, p. 223–229, 2022.

FARRELL, PM; WHITE, TB; REN, CL; HEMPSTEAD, SE; ACCURSO, F.; DERICHS, N.;

Year VI, v.1 2026 | Submission: 01/16/2026 | Accepted: 01/18/2026 | Publication: 01/20/2026

HOWENSTINE, M.; MCCOLLEY, SA; ROCK, M.; ROSENFELD, M.; SERMET-GAUDELUS, I.; SOUTHERN, KW; MARSHALL, BC; SOSNAY, PR *Diagnosis of cystic fibrosis: consensus guidelines from the Cystic Fibrosis Foundation.* Journal of Pediatrics, vol. 181, suppl., p. S4–S15.e1, 2017.

FRASÃO, G. *Cystic fibrosis can be identified in the first days of a baby's life.* Brasília: Ministry of Health, 2021.

Oswaldo Cruz Foundation. Fernandes Figueira National Institute of Women's, Children's and Adolescents' Health. *Key questions about cystic fibrosis: how to diagnose it?* Rio de Janeiro, 2023.

GABATZ, RIB; RITTER, NR. *Children hospitalized with cystic fibrosis: perceptions about multiple hospitalizations.* Brazilian Journal of Nursing, v. 60, n. 1, p. 37–41, 2007.

GRAY, Henry FRS *Gray's anatomy: the anatomical basis of clinical practice.* 40th ed. Rio de Janeiro: Elsevier, 2010.

HAACK, A.; ARGÃO, GC; NOVAES, M. *Pathophysiology of cystic fibrosis and drugs commonly used in respiratory manifestations: what we should know.* Communications in Health Sciences, v. 25, n. 3/4, p. 246–262, 2014.

MATOS, Bruna de Almeida; MARTINS, Rita Cristina. *Cystic fibrosis: a literature review.*
Brazilian Journal of Surgery and Clinical Research, v. 29, no. 2, p. 114–119, 2019.

MOORE, Keith L.; DALLEY, Arthur F.; AGUR, Anne MR . *Clinically Oriented Anatomy.* 8th ed. Rio de Janeiro: Guanabara Koogan, 2019.

ORLANDIN, Caisa Brunelli. *Project for a physical and pulmonary rehabilitation protocol in adult patients with cystic fibrosis with severe respiratory disease.* 2019. Thesis (Technical Project) – São Paulo State Health Secretariat, Ribeirão Preto, 2019.

ROSA, FR; DIAS, FG; NOBRE, LN; MORAIS, HA *Cystic fibrosis: a clinical and nutritional approach.* Revista de Nutrição, v. 21, n. 6, p. 725–737, 2008.

ROSENSTEIN, BJ. *Cystic fibrosis.* MSD Manual, 2024.

SCHNEIDERS, L. *Cystic fibrosis is genetic and more common in childhood.* Brasília: Ministry of Health, 2024.

SNELL, G.; REED, A.; STERN, M.; HADJILIADIS, D. *The evolution of lung transplantation for cystic fibrosis: a 2017 update.* Journal of Cystic Fibrosis, vol. 16, no. 5, p. 553–564, 2017.