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Cystic Fibrosis

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What is Cystic Fibrosis (CF)?

- CF is an autosomal recessive inherited disease (McCance et al., 2019).
- Result of dysfunctional epithelial chloride ion transport (McCance et al., 2019).
- Mainly diagnosed in infancy (Turcios, 2020).
- More than 75 percent of people with CF are diagnosed by age 2 (Cystic Fibrosis Foundation, n.d.).

Why Cystic Fibrosis?

- Affects 1 in every 3500 Caucasian births (Karrar et al., 2022).
- Affects approximately 30,000 people in the U.S. and 80,000 worldwide (Karrar et al., 2022).
- Patients require support from healthcare services from diagnosis onwards (Keogh et al., 2017).
- Although treatment has become more targeted and the average life expectancy has increased in recent years, no cure has been discovered (Keogh et al., 2017).
- The cause of death is almost always respiratory failure. (Keogh et al., 2017).
- Multisystem involvement results in a frequent requirement of anesthesia for surgical procedures (May et al., 2019).

Diagnostic Testing

- Begins with a positive newborn screening, most commonly due to abnormal levels of pancreatic enzyme precursor immunoreactive trypsinogen (Trucios, 2020).
- Definitive diagnostic tool is Sweat Testing Technique (Karrar et al., 2022).
- Measurement of the chloride and sodium concentration of sweat (Karrar et al., 2022).
- Usually performed 48 hours after birth (Turcios, 2020).
- Chloride levels begin to increase 24 hours after birth (Karrar et al., 2022).
- A very high concentration of chloride is diagnostic for Cystic Fibrosis (Karrar et al., 2022).

Pathophysiological Processes Signs & Symptoms

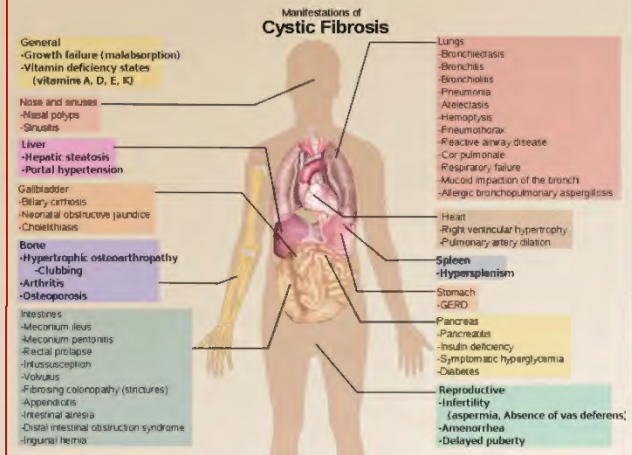

Figure 1. Clinical Manifestations of Cystic Fibrosis *(Mansour, 2022).*

- Often, the earliest symptoms are GI and nutritional disorders (Turcios, 2020).
- 15% of affected infants present with meconium ileus at birth (Turcios, 2020).
- Non-pulmonary indicators for screening include electrolyte imbalance, rectal prolapse, diabetes mellitus, recurrent pancreatitis, digital clubbing, chronic sinusitis, nasal polyps, and biliary cirrhosis (Turcios, 2020).

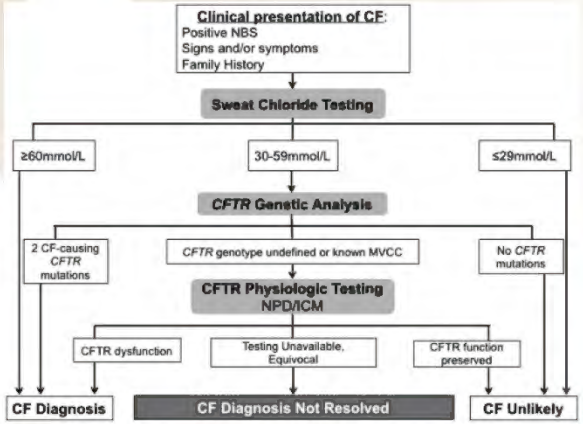

Figure 2. Diagnostic Pathway of Cystic Fibrosis *(Farrell et al., 2017).*

Underlying Pathophysiology

- Result of a gene mutation that causes the abnormal production of cystic fibrosis transmembrane conductance regulator (CFTCR) protein (McCance et al., 2019).
- CFTCR protein is an activated chloride channel on the surface of numerous epithelial cells (Karrar et al., 2022).
- CFTCR protein regulates the movement of chloride ions and water secretion and absorption (de Faria Poloni et al., 2021).
- Channel dysfunction results in ionic imbalance, causing the secretion of thick and dehydrated mucus and fat malabsorption (de Faria Poloni et al., 2021).
- Abnormal epithelial fluid absorption in the lungs raises the osmotic pressure in the mucus layer compared to the periciliary layer causing hyper-concentrated mucus build-up in small airways (Turcios, 2020).

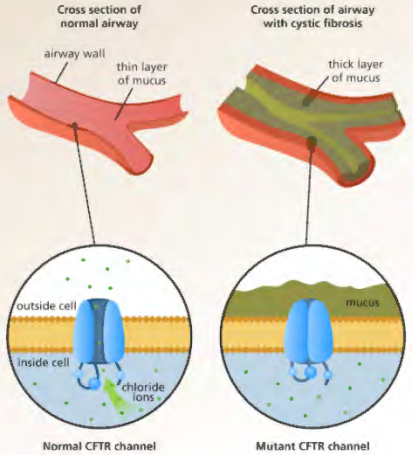

Figure 3. *Pathologic process of Cystic Fibrosis (YourGenome, 2021).*

Significance of Pathophysiology

- Viscous secretions contribute to mucus plugging, poor secretion mobilization, and possibly eventual respiratory failure (May et al., 2019).
- Increased accumulation of mucus and decreased mucociliary clearance facilitate colonization by organisms, especially bacteria, resulting in recurrent respiratory infections (Karrar et al., 2022).
- The most common bacterial colonizations are pseudomonas, Haemophilus influenza, and staphylococcus aureus (Karrar et al., 2022).
- Endobronchial infection induces a hypersensitive inflammatory response characterized by elevated levels of neutrophil elastase in the airway that contribute to excess mucus adhesion and tenacity (Turcios, 2020).
- An exaggerated inflammatory response can result in damage to the structural integrity of the airways and the development of irreversible abnormal dilatation of affected segmental and subsegmental bronchi (Turcios, 2020).
- CF lung disease is progressive, and dysfunction leads to increased work of breathing, impaired gas exchange progressing to arterial hypoxemia, and eventually respiratory failure (Turcios, 2020).
- Destruction of acinar pancreatic tissue, lack of enzyme activity, and pancreatic duct obstruction result in malabsorption of fats and proteins (Turcios, 2020).

Treatment

Symptoms and severity vary greatly from person to person; therefore, treatment plans are highly individualized (Cystic Fibrosis Foundation, n.d.).

As there is no cure, treatment for CF centers around symptom management (Keogh et al., 2017).

According to the Cystic Fibrosis Foundation (n.d.), the following treatments are most common:

- Chest physiotherapy to loosen and clear mucus accumulation in the airway.
- Bronchodilators to maximize airflow into the lungs.
- Mucolytic agents and hypertonic saline nebulizers to thin secretions.
- Antibiotics to treat chronic lung infections.
- Steroids to reduce airway inflammation.
- Pancreatic enzyme supplementation with all meals and snacks to improve nutrient absorption.
- Pulmonary strength training
- CFTR modulators can be given in cases of specific gene mutations.

If respiratory symptoms become unmanageable, a lung transplant may be required to improve quality of life (Turcios, 2020).

Implications for Nursing Care

- Symptoms such as pancreatic enzyme deficiency, reduced nutritional absorption, increased work of breathing and inadequate nutrient intake all contribute to poor growth, suboptimal clinical outcomes, and reduced life expectancy (McCance et al., 2019).
- Oral pancreatic enzyme replacement with every meal and snack is crucial in maintaining nutritional status (McCance et al., 2019).
- Infection prevention is paramount to decreasing the incidence of lung infections, preserving healthy lung tissue, and maintaining airway integrity (Turcios, 2020).
- Knowledge of the pathophysiologic process could result in more appropriate intervention selection for managing symptoms such as acute respiratory distress.

Considerations for Anesthesia

- The extent of pulmonary involvement is the primary determinant of appropriateness for surgery (Elisha et al., 2023).
- Patients should be evaluated several days prior to scheduled surgery, and patients experiencing symptomatic respiratory distress should be treated in an inpatient setting where appropriate hydration and respiratory management can be administered (Elisha et al., 2023).
- Patients who are appropriate for surgery may still present complications for perioperative care, including alterations in ventilation-perfusion ratios, intrapulmonary shunting, mucous plugging, and increased dead space (May et al., 2019).

Conclusions

- Cystic Fibrosis is a complex disease process caused by a gene mutation resulting in defective chloride channel transport (Turcios, 2020).
- CF affects a wide variety of body systems (Mansour, 2022).
- There is no cure for CF (Keogh et al., 2019).
- Treatment is multimodal and progressive (Cystic Fibrosis Foundation, n.d.).
- Preventing respiratory infections is crucial to preserving lung functionality (Karrar et al., 2022).
- Management of patients with CF would improve with increased understanding of the pathophysiological process and commitment to diligent infection prevention from healthcare professionals.

References

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