

Review Article

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Impact of cystic fibrosis on pediatric oral health: a comprehensive review

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ABSTRACT

Cystic fibrosis (CF) is a complex genetic disorder that affects multiple systems, including the oral cavity. In children, CF leads to unique challenges in maintaining oral health due to the interplay of systemic factors, dietary requirements, and medical treatments. Alterations in salivary flow and composition, often observed in CF patients, contribute to an increased risk of xerostomia, enamel erosion, and periodontal disease. These issues are further exacerbated by high-calorie, carbohydrate-rich diets prescribed to manage metabolic needs, creating a favorable environment for caries development. The use of inhaled corticosteroids, nebulized antibiotics, and other essential medications introduces additional oral health challenges, such as fungal infections and changes in the oral microbiota. Gastroesophageal reflux disease, a common comorbidity in CF, contributes to dental erosion through repeated exposure to gastric acids. Despite these risks, frequent use of antibiotics in CF patients has shown a paradoxical reduction in dental caries prevalence, highlighting the complex oral microbial dynamics in this population. Preventive strategies include dietary counseling, regular fluoride application, and tailored oral hygiene routines, such as the use of electric toothbrushes and interdental brushes. Therapeutic interventions focus on minimally invasive techniques, fluoride-releasing restorative materials, and the application of sealants to protect vulnerable tooth surfaces. Collaboration among dentists, pediatricians, dietitians, and CF specialists is crucial to address the multifactorial challenges and improve both oral and systemic outcomes. The relationship between cystic fibrosis and oral health underscores the importance of integrating dental care into the broader management of CF. Early identification of oral complications and the implementation of personalized prevention and treatment strategies can significantly enhance the quality of life for pediatric CF patients while reducing the long-term burden of oral diseases.

Keywords: Cystic fibrosis, Pediatric oral health, Enamel erosion, Preventive dentistry, Salivary dysfunction

INTRODUCTION

Cystic fibrosis (CF) is a complex, autosomal recessive genetic disorder primarily affecting the respiratory and gastrointestinal systems. It arises from mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene, which leads to defective chloride ion transport and the production of thick, viscous secretions in multiple organ systems.¹ While the respiratory and digestive implications of CF are well-documented, its impact on oral health is increasingly recognized as an important area of study.

Children with CF often face unique oral health challenges. These issues stem from systemic factors such as chronic malnutrition, long-term antibiotic use, and altered salivary flow. These systemic conditions can predispose pediatric patients to dental caries, enamel hypoplasia, and other oral manifestations.² In addition, CF patients often require a high-calorie diet rich in carbohydrates to combat their metabolic demands, inadvertently increasing their risk of oral diseases, including caries and gingivitis.³

Medications used to manage CF also play a critical role in oral health. Nebulized antibiotics, inhaled corticosteroids, and pancreatic enzyme supplements may directly or indirectly contribute to xerostomia and dental erosion.⁴ Additionally, gastroesophageal reflux disease (GERD), common in CF, exacerbates dental enamel erosion due to chronic acid exposure. From a microbial perspective, the oral cavity of CF patients exhibits altered microbiota, often characterized by increased colonization of opportunistic pathogens, such as *Pseudomonas aeruginosa* and *Candida albicans*.³ These shifts in oral microbial communities can exacerbate inflammation and contribute to periodontal disease.

Despite these challenges, studies suggest that children with CF may have a lower prevalence of dental caries than their healthy counterparts, potentially due to frequent antibiotic use and altered salivary composition.² However, this advantage does not eliminate their susceptibility to other oral health complications. Moreover, the quality of life for children with CF can be significantly impacted by their oral health status, underscoring the need for a multidisciplinary approach to care. This review aims to provide a comprehensive overview of the impact of cystic fibrosis on pediatric oral health, exploring its pathophysiological mechanisms, clinical manifestations, and potential preventive and therapeutic strategies.

REVIEW

CF significantly impacts pediatric oral health due to its systemic effects and treatment requirements. One prominent issue is the alteration in salivary composition, which reduces its protective effects. Saliva in children with CF often shows elevated levels of calcium and phosphorus, contributing to calculus formation while reducing caries risk. However, this is offset by the increased prevalence of

enamel hypoplasia, which makes teeth more susceptible to mechanical and chemical damage.⁴ The use of high-calorie, carbohydrate-rich diets recommended for CF patients further exacerbates the risk of dental decay, despite the potential benefits of frequent antibiotic use.

The oral microbiota in CF patients also undergoes significant changes, often reflecting shifts seen in respiratory infections. Colonization by pathogens such as *Pseudomonas aeruginosa* and *Candida albicans* can contribute to oral health complications, including periodontal disease and mucosal infections. These pathogens are thought to originate from the shared microbial environment of the respiratory and oral cavities, highlighting the interconnected nature of these systems.⁵ The frequent use of inhaled corticosteroids and nebulized medications further complicates oral health by contributing to xerostomia and soft tissue inflammation. Addressing these multifaceted issues requires tailored preventive strategies to mitigate both local and systemic risk factors.

Pathophysiological link between cystic fibrosis and oral health in pediatric patients

CF is a systemic condition primarily characterized by defective chloride ion transport caused by mutations in the CFTR gene. This dysfunction leads to a cascade of physiological alterations that extend beyond the respiratory and gastrointestinal systems to include significant impacts on oral health. The oral manifestations of CF in pediatric patients reflect the interplay between systemic deficiencies, microbial imbalances, and chronic medication use. The salivary composition in CF patients is notably altered, often showing increased viscosity and decreased buffering capacity. These changes can impair the protective functions of saliva, such as its ability to neutralize acids and inhibit microbial colonization. Reduced salivary flow has also been associated with a higher prevalence of dental caries and mucosal infections. The chemical composition of saliva in CF patients frequently includes elevated levels of calcium and phosphate, promoting the rapid deposition of dental calculus while reducing caries incidence in some cases.⁶ Additionally, chronic nutritional deficiencies in CF patients, particularly in fat-soluble vitamins like A, D, E, and K, have repercussions for oral health. Vitamin D deficiency, for example, has been linked to compromised mineralization of dental enamel, increasing the risk of enamel hypoplasia and erosion. Malabsorption of other essential nutrients further exacerbates oral complications by impairing tissue repair and immune responses within the oral cavity.⁷

Microbial dysbiosis is a hallmark of CF-related oral health issues. The oral microbiome in these patients often mirrors the alterations seen in their respiratory tracts, with increased prevalence of pathogens such as *Pseudomonas aeruginosa* and *Candida albicans*. These organisms can contribute to chronic periodontal inflammation and

mucosal infections. Research highlights that the disrupted microbial environment in the oral cavity may facilitate the establishment of biofilms, which are more resistant to traditional antimicrobial treatments.⁸ Moreover, frequent medication use plays a critical role in shaping the oral health outcomes of pediatric CF patients. Inhaled corticosteroids, while vital for managing respiratory symptoms, have been associated with the development of oral candidiasis. Nebulized antibiotics, similarly, may inadvertently encourage antibiotic-resistant oral flora. Additionally, the systemic use of pancreatic enzyme supplements, which are often mixed with carbohydrate-rich foods, can heighten the risk of caries in these children.⁹ Furthermore, the link between GERD and oral health in CF patients is another critical area of concern. GERD, a common complication of CF, often leads to acid reflux that contributes to dental erosion and sensitivity. The constant exposure to gastric acids not only damages the enamel but also disrupts the oral pH balance, favouring the growth of acid-tolerant bacteria and further worsening dental conditions.¹⁰

Compromised immune function in CF patients, compounded by recurrent infections and inflammation, further complicates their oral health status. Poor wound healing, coupled with frequent oral surgeries or dental procedures, increases the risk of infections and delays recovery times. The systemic inflammation characteristic of CF is also reflected in heightened periodontal inflammation, which can lead to early-onset gingivitis or periodontitis in pediatric patients.¹¹ Addressing the oral health challenges in children with CF necessitates a multidisciplinary approach. Regular dental assessments, combined with tailored preventive strategies, are critical in mitigating the compounded effects of systemic and local factors. Enhancing awareness among caregivers and healthcare providers regarding the bidirectional relationship between CF and oral health is essential for improving overall patient outcomes.

Oral manifestations and common dental complications in children with cystic fibrosis

CF presents a spectrum of oral health challenges that are directly linked to the systemic implications of the disease and its associated treatments. Pediatric patients with CF often exhibit unique oral manifestations, which include a combination of salivary gland dysfunction, enamel hypoplasia, and increased susceptibility to periodontal diseases. Salivary alterations, such as decreased flow rates and increased viscosity, have been observed, reducing saliva's natural cleansing and buffering capacity. This contributes to the retention of food particles and microbial overgrowth, compounding the risk of dental plaque accumulation and gingival inflammation.¹²

A notable dental complication among CF patients is enamel hypoplasia, which is believed to arise from systemic metabolic disturbances during tooth development (Figure 1). This condition results in enamel that is thinner

and more prone to wear, erosion, and discoloration. The compromised enamel integrity increases the likelihood of dental caries and sensitivity, particularly when exacerbated by dietary habits necessitated by the high-calorie nutritional requirements of CF patients.¹³ Additionally, the frequent consumption of carbohydrate-rich foods and sugary supplements to maintain energy levels inadvertently fuels cariogenic bacteria, further amplifying caries risk.



Figure 1: Child with enamel hypoplasia.¹⁴

Periodontal health is another critical concern in children with CF. Studies indicate a heightened prevalence of gingivitis and early-onset periodontitis in this population, often linked to their altered oral microbiome. The chronic use of antibiotics, while effective in managing respiratory infections, disrupts the balance of normal oral flora. This alteration can lead to overgrowth of opportunistic pathogens such as *Candida albicans* and *Pseudomonas aeruginosa*, both of which have been implicated in periodontal tissue damage.¹⁵ Dental erosion, frequently seen in CF patients, is often attributed to GERD, a common comorbidity in CF. Acidic reflux into the oral cavity repeatedly exposes the teeth to low pH levels, which demineralizes enamel and increases its susceptibility to physical and chemical wear. The cumulative effect of enamel demineralization and erosion creates a conducive environment for the rapid progression of dental decay.¹⁶

Another significant factor impacting oral health in children with CF is the use of inhaled corticosteroids and nebulized medications. These treatments, essential for managing pulmonary symptoms, have been associated with oral dryness and irritation, making the mucosal tissues more vulnerable to candidiasis and other fungal infections. Xerostomia, or dry mouth, not only impairs the self-cleaning mechanisms of saliva but also fosters bacterial growth, leading to plaque formation and subsequent gingival inflammation.¹⁷

The impact of oral health extends beyond the physical manifestations to affect the overall quality of life of CF patients. Pain, difficulty eating, and aesthetic concerns caused by dental complications can exacerbate the psychological burden of living with a chronic disease. This highlights the importance of integrating dental care into the

multidisciplinary management of CF. Regular dental visits, fluoride treatments, and preventive education tailored to the specific needs of CF patients are pivotal in mitigating these challenges.¹⁸

Preventive strategies and therapeutic interventions for improving oral health outcomes

Managing the oral health of pediatric CF patients is challenging due to the systemic complications of the disease and its associated treatments. A strategic approach combining preventive care, therapeutic interventions, and multidisciplinary collaboration is essential to address their unique oral health needs effectively. By focusing on personalized care plans, dental professionals can significantly mitigate oral health risks and improve overall outcomes. Dietary modifications are among the first steps in preventive care for CF patients. These children often follow high-calorie diets to meet their metabolic demands, but these diets frequently include sugary and carbohydrate-rich foods that elevate the risk of dental caries. Counseling caregivers to incorporate tooth-friendly snacks such as cheese, nuts, and raw vegetables can counterbalance this risk. The use of xylitol-containing products, including chewing gums and mints, has demonstrated efficacy in reducing bacterial colonization and promoting remineralization. Xylitol disrupts the energy production processes in *Streptococcus mutans*, a leading contributor to dental caries, thereby reducing its growth and acid production.¹⁹

Salivary gland dysfunction is a prevalent issue among CF patients, contributing to xerostomia and the resultant increased risk of caries, gingivitis, and mucosal lesions. Addressing salivary deficiencies involves recommending sugar-free lozenges or gums to stimulate saliva production. In cases of severe xerostomia, artificial saliva substitutes containing enzymes and electrolytes can restore moisture and enhance the natural protective barrier of the oral cavity. The regular use of fluoride varnishes, mouth rinses, or prescription-strength toothpaste is a crucial component of preventive care, as fluoride strengthens enamel and reduces its susceptibility to demineralization. Research has shown that fluoride treatments significantly decrease the progression of early carious lesions, especially in high-risk populations.²⁰

Plaque control and the prevention of periodontal disease require meticulous oral hygiene practices. Daily brushing with fluoridated toothpaste, complemented by the use of interdental brushes or floss, can remove plaque from hard-to-reach areas. Electric toothbrushes are particularly beneficial for children with limited manual dexterity. For professional care, dental prophylaxis every three to six months is recommended to prevent the build-up of plaque and calculus. Chlorhexidine mouth rinses have proven effective in reducing gingival inflammation and controlling bacterial load, offering additional protection against periodontal disease. However, long-term use of

chlorhexidine must be monitored to avoid potential side effects, such as tooth staining and taste alteration.²¹

The use of antibiotics and corticosteroids to manage respiratory symptoms in CF patients introduces additional oral health challenges. Prolonged use of these medications alters the oral microbiota, often leading to an overgrowth of opportunistic pathogens such as *Candida albicans*. Targeted antifungal therapies, including nystatin or fluconazole, are effective in treating oral candidiasis. To prevent recurrent fungal infections, patients should rinse their mouths thoroughly after using inhalers and nebulizers. Additionally, inhaler spacers can reduce the deposition of medication on oral tissues, minimizing the risk of irritation and fungal colonization.²²

Therapeutic interventions focus on repairing and restoring damage caused by systemic conditions and poor oral health. The use of dental sealants is a highly effective measure for protecting occlusal surfaces of molars and premolars from decay. Sealants act as a physical barrier, preventing food particles and bacteria from settling in pits and fissures. Restorative treatments for carious lesions should prioritize materials with fluoride-releasing properties, such as glass ionomer cements and resin composites, as these materials provide long-term enamel protection. Additionally, minimally invasive techniques such as atraumatic restorative treatment are ideal for pediatric patients with CF, as they reduce discomfort and stress during dental procedures.²³

Collaboration among healthcare providers is essential for successful oral health management in CF patients. Dentists, pediatricians, and dietitians must coordinate care to address the multifactorial aspects of CF. For example, addressing the nutritional requirements while simultaneously mitigating cariogenic risks requires a delicate balance that benefits from interprofessional input. Regular dental assessments should be synchronized with routine medical checkups to monitor and address emerging oral health issues promptly.²⁴ Education plays a pivotal role in preventive care. Caregivers should be equipped with knowledge about CF-specific oral health risks and trained in implementing effective oral hygiene routines. Simple techniques, such as the use of visual aids to demonstrate proper brushing and flossing, can empower families to manage oral health more effectively at home. Additionally, caregivers should be encouraged to schedule regular dental visits, ensuring early detection and management of oral health problems before they escalate. Preventive care and therapeutic interventions tailored to the unique challenges faced by CF patients can significantly improve their oral health outcomes, reducing the overall burden of disease and enhancing their quality of life.

CONCLUSION

Managing oral health in children with cystic fibrosis requires a holistic approach that addresses systemic,

dietary, and microbial factors. Proactive preventive care, including tailored dietary advice, fluoride application, and professional cleanings, is essential to mitigate oral health risks. Therapeutic interventions should prioritize minimally invasive and fluoride-releasing materials to protect and restore oral structures. A multidisciplinary collaboration between healthcare providers ensures comprehensive care, enhancing both oral and overall health outcomes for CF patients.

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