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Authors	Coffey, Niamh;O'Leary, Fiona;Burke, Francis M.;Hayes, Martina
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Oral Disease in People with Cystic Fibrosis

N. Coffey, F. O' Leary, F.M. Burke, M. Hayes

Cork University Dental School and Hospital, University College Cork, Ireland.

Introduction

Cystic Fibrosis (CF) is a well-characterized, severe monogenic recessive disorder. It is mainly found in white populations of European ancestry and arises from mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene on chromosome 7. It is a multi-organ disease with manifestations primarily seen in the pulmonary, digestive, and reproductive systems. The median age of diagnosis in Ireland is 0.29 years [1, 2]. Ireland has the highest incidence of CF in the world. Currently, there are approximately 1380 People With CF (PWCF) in Ireland[2]. The CF population worldwide is seeing an increase in life expectancy. In Ireland, approximately 58.5% of the CF patient population in Ireland is aged 18 years or older and the median age of survival is 44.4 years, an increase from 36.6 years in 2008 [2].

Lung Transplantation may be carried out in cases of severe lung disease. Between 2011 and 2018, 68 PWCF received a bilateral lung transplant in the Mater Misericordiae University Hospital, Dublin [2]. People undergoing lung transplantation are often recommeded to be declared "dentally fit" pre-operatively, but there is currently no clear evidence-based guidelines on the dental management of PWCF in general or, specifically, pre- or post- transplantation.

Dental diseases are extremely prevalent worldwide and can significantly impact on a person's quality of life[3]. The most common oral diseases are dental caries (tooth decay), periodontal (gum) disease, developmental defects of enamel and candida infection.

Research has shown evidence of a relationship between dental disease and a number of systemic diseases. Studies have shown that there is greater prevalence and severity of periodontal disease in diabetic subjects than in nondiabetic subjects [4]. As 19.6% of PWCF in Ireland suffer from Cystic Fibrosis Related Diabetes (CFRD) [2], it is possible that this subset of PWCF are at higher risk of periodontal disease. Furthermore, it has been found that there is worsening glycemic control in diabetic patients with periodontal disease compared with those without suggesting a bidirectional relationship. Diabetic subjects who also have periodontal disease have been found to be at a three times greater risk of diabetic complications. There are also strong links between periodontal disease and cardiovascular disease and the development of atherosclerosis. Perhaps of most importance to PWCF is the risk that the oral cavity can ask as a reservoir of respiratory pathogens, which can result in pulmonary disease and pneumonia [4]. Due to the increasing life expectancy of people with CF, it is timely that we review the literature available regarding any potential links between Cystic Fibrosis and oral diseases.

What is the evidence?

Dental Caries

Dental caries is the localised dissolution of dental hard tissues (enamel and dentine) by acidic by-products from the bacterial fermentation of sucrose and other dietary carbohydrates. It has been hypothesised that individuals with CF are at higher risk of dental caries due to certain risk factors. These include Gastro-Oesophageal Reflux Disease (GORD), high levels of *Streptococcus mutans*, high calorie diets to avoid malnutrition, and frequent prescription of sugar containing antibiotics [5, 6].

Despite these risk factors, studies have shown that children with CF are actually at lower risk of developing dental caries. Two systematic reviews [6, 7] showed that, in general, there was equal or lesser caries risk in children with CF compared to a control group. It is hypothesised [6, 8] that this is due to long term antibiotics that PWCF must take for recurrent respiratory infections. These antibiotics (including penicillins) can target caries-causing bacteria such as *S. mutans*. Another hypothesis is that PWCF may be more health conscious and therefore may engage in more meticulous oral hygiene regimes- which will reduce the risk of caries (and also the risk of periodontal disease)[6]. Yet another hypothesis put forward is that supplemental Pancreatic Enzyme Replacement Therapy (PERT) could potentially reduce the incidence of caries due to the inhibition of plaque accumulation [8]. PERT is currently prescribed to at least 89.9% of PWCF in Ireland [2].

On the other hand, despite this seemingly protective effect CF bestows on individuals during childhood, Chi remarked that "adolescents with CF may not be at lower caries levels than those without CF". Similarly, Dabrowska et al (2006) found that there was a higher incidence of caries in CF patients between the ages of 6-12 than a control group [5]. There are a number of reasons put forward for this: caries risk tends to increase during adolescence due to behavioural changes such as decreased frequency of toothbrushing and poor diet, and the antibiotics used are changed to address the fact that *pseudomonas aeruginosa* becomes the most prevalent pulmonary pathogen - these antibiotics (such as tobramycin) do not target *S. mutans*.

Periodontal Disease

Periodontal disease can be broadly defined as the "chronic inflammatory conditions that affect the tissues surrounding and supporting the teeth". The main cause of the inflammatory condition is poor oral hygiene which can lead to an accumulation of pathogenic microbial biofilm, or plaque, at and below the gingival margin [3]. The initial presentation is that of gingivitis, red inflamed gums that bleed on brushing. In individuals where there is dysbiosis or immune overreaction of the host to microbial presence, gingivitis may progress to periodontitis, which results in damage to the alveolar bone (the supporting bone around the teeth). The final result of this process can be tooth loss. Due to recurrent respiratory infections, people with CF often breathe through the mouth- this can promote malocclusion/misalignment of teeth which may predispose to periodontitis [5].

There are mixed findings regarding periodontal health in PWCF. Some studies concluded that there were lower levels of gingivitis in the CF group compared to a control group or to the national average [9-11]. One study [12] found that a particular subset of patients (those with CF aged between 6 and 9.5 years) had higher levels of gingivitis. A number of studies, including those by Aps, Van Maele and Martens in 2001/2002 found there was no significant difference in oral hygiene between PWCF and controls [13, 14]. A number of studies showed that PWCF had higher levels of dental calculus compared to a control group [8, 9, 12]. Only one study looked at periodontal pocketing depths- a major determinant of periodontal disease.

Interestingly, this study found that although plaque levels were generally "moderate or severe", there was no patient with severe gingivitis or periodontal pocketing depths greater than 4mm, i.e. no sign of clinical periodontal disease [15]. This study hypothesised that the reduced levels of clinical periodontitis could be due to frequent intake of systemic antibiotics.

Developmental defects of enamel (DDEs)

Developmental defects of enamel (DDEs) are commonly encountered in general dental practice where they may present as enamel hypoplasia or hypomineralization. The main clinical problems arising are compromised aesthetics, tooth sensitivity and increased risk for dental caries and tooth wear [16].



Figure 1: example of enamel hypoplasia-a type of DDE. Picture copyright Dr. Mairead Harding, Cork University Dental School and Hospital, from FACCT Study CARG/HRB2012/34

The majority of studies [10-12, 14] showed that there was a higher incidence of DDEs in the CF population compared to the healthy population.

Candida

Among the factors that predispose individuals to oral candidiasis are use of broad- spectrum antibiotics and corticosteroids [17]. Approximately 62% of PWCF in Ireland have taken long term (>3months) oral antibiotics in the last year, and approximately 29% have taken inhaled steroids [2]. Oral candidiasis can cause frequent and significant oral discomfort, pain, altered taste sensation (dysgeusia), and aversion to food [17].

Discussion

The evidence is mixed as to whether or not PWCF are at greater risk of dental disease. There are limitations to some previous studies including small sample sizes, the young age of the population studied and the age of the studies. Most were carried out before the introduction of CFTR modulators so the impact –if any- of these on the oral environment is yet to be researched.

The population of PWCF is increasing, as is their life expectancy. As the majority of PWCF in Ireland are now adults, it would be beneficial to conduct a study to look at the oral health of these individuals.

If they are found to be at higher risk of dental disease, then it would be prudent to categorise them as "high risk" from the moment of diagnosis and make provision for them as "people needing special care" as outlined in the National Oral Health Policy, "Smile agus Sláinte". It may also be wise to include a dentist as part of the multi-disciplinary team involved in the care of PWCF- including working with nutritionists when formulating a diet plan. Furthermore, people undergoing lung transplants often require an oral health clearance, but the specifics of this have not been clearly defined. There is potentially need for communication between the dental team and CF/respiratory or surgical team, and to create guidelines for the oral care of people with CF.

Conclusion

There is a need for good quality studies into the oral health of individuals, specifically adults, with Cystic Fibrosis. There is currently insufficient evidence to determine the impact of CF on oral health but PWCF should be encouraged to attend their dentists regularly to maintain good oral health and minimise any future complications.

Declaration of Conflicts of Interest:
The authors declare that they have no conflict of interest.

Corresponding Author:
Niamh Coffey,
Restorative Dentistry,
Cork University Dental School and Hospital,
University College Cork,
Ireland,
T12 E8YV.
Email: niamh.coffey@ucc.ie

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