Disease/Medical Condition

CYSTIC FIBROSIS
(also known as CF)

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Is the initiation of non-invasive dental hygiene procedures* contra-indicated? No

- Is medical consult advised? No, assuming CF is well controlled and there are no concerns with respiratory status.

Is the initiation of invasive dental hygiene procedures contra-indicated?** No

- Is medical consult advised? See above.
- Is medical clearance required? No, not typically. However, if respiratory function is suspected to be severely compromised and/or unstable, medical clearance should be obtained. Also, persons who have undergone lung, heart, and/or liver transplants will be on immunosuppressive medications to reduce organ rejection, and such patients/clients should be medically cleared. Similarly, prolonged use and/or high doses of systemic (oral) corticosteroids may predispose to infection, and medical clearance (and potential dose adjustment) should be sought in these circumstances.
- Is antibiotic prophylaxis required? No, not typically. However, antibiotic prophylaxis should be considered for patients/clients who are taking medications that induce immunosuppression (e.g., immunosuppressants for lung, heart, and liver transplants, as well as prolonged use and/or high doses of systemic steroids).
- Is postponing treatment advised? No, if CF is well controlled. Yes, if the patient/client is acutely ill or has respiratory compromise (or other co-morbidities) incompatible with the delivery of dental hygiene care; routine dental hygiene treatment should be postponed until resolution of the acute illness or better CF respiratory control is achieved.

Oral management implications

- The dental hygienist can play an important role in promoting health in patients/clients with CF. Disease control, nutritional status, and status of the oral care regimen should be routinely assessed.
- Patients/clients with CF typically have medical evaluations every several months. Therefore, the patient/client’s medical history should be regularly updated, including questions regarding lung function, hospitalization, potential contraindications to dental hygiene care, and possible need for medical consult prior to continuing care delivery.
- Patients/clients with CF typically follow a high-fat, high-carbohydrate diet, which can promote tooth decay. Prophylaxis with topical fluoride treatment and sealants should be considered, along with referral to registered dietitian to promote dietary choices that support oral health.
- Use of sodium bicarbonate rinses or xylitol after consuming sugary drinks or snacks, and after using an inhaler/nebulizer, should be encouraged.
- Appointment scheduling may need to be flexible, because patients/clients with CF may spend several hours per day undergoing breathing/respiratory therapy and keeping other health-related appointments.
- Chair position should be semi-supine or upright to help improve the patient/client’s breathing and respiratory comfort; the upright sitting position aids in the clearance of secretions from the bronchi and trachea via coughing. Short appointments are indicated if there is significant compromise of lung function.
- Persons with a known respiratory risk or chronic pulmonary disorders, such as CF, can potentially aspirate septic material or pathogenic microorganisms from biofilm associated with periodontal inflammatory disease, and should not be treated with ultrasonic scalers.

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Oral management implications (con’t)

■ While many persons with CF have a chronic cough and may appear to be ill, they are not contagious to the dental hygienist or other patients/clients, unless they have signs/symptoms of acute upper respiratory tract infection with fever and coryza.\(^1\)

■ While CF is not an immunosuppressive condition (unless there is associated drug-mediated immunosuppression associated with transplantation), persons with CF, given their compromised respiratory function, should have their exposure minimized to others who have contagious respiratory illnesses such as influenza or the common cold.

■ A plan to assess alveolar bone health should be implemented due to the increased risk of osteoporosis.

Oral manifestations

■ Cystic fibrosis does not usually affect teeth directly, although poor nutrition may affect tooth growth and structure. As well, high calorie foods consumed by CF patients/clients often contain a lot of sugar, which, while beneficial for the treatment of CF, may promote dental caries.

■ Enamel defects are common, including demarcated opacities, diffuse opacities, and enamel hypoplasia. These can be disfiguring.

■ There are conflicting studies regarding plaque levels in persons with CF, with more recent studies showing no difference from the general population (while older studies reported decreased plaque).

■ Persons with CF may be at reduced risk of periodontal disease (possibly related to certain antibiotic – e.g., azithromycin – penetration of the periodontal tissues and retention in the periodontal pocket).

■ Patients/clients with CF tend to experience fewer gingival bleeding sites (possibly related to long-term antibiotic regimens) than persons without CF.

■ Oral candidiasis occurs at elevated rates, due to long-term antibiotic regimens and the use of inhaled steroids, as well as from the effects of CF-related diabetes.

■ CF affects all exocrine glands, including the salivary glands. However, the parotid glands are virtually pure serous (rather than mucus-secreting) glands, and thus their saliva is only minimally affected. Submandibular, sublingual, and minor salivary glands, by contrast, show greater changes in gland architecture and saliva composition. As well, the submandibular glands are usually enlarged and easily palpable.

■ Mouth breathing and anterior open bite are often seen in patients/clients with CF, given the association with chronic nasal and sinus obstruction.

■ Some medications used to manage CF — such as bronchodilators and corticosteroids — can contribute to oral disease. \(\beta_2\) (beta2) agonist inhalers (i.e., bronchodilators such as salbutamol) reduce salivary flow, resulting in xerostomia. In addition, \(\beta_2\) agonist inhalers also lower plaque pH, cause unpleasant taste sensation, and are associated with increased prevalence of dental caries and gingivitis in patients/clients who frequently use them. Oral candidiasis, gingivitis, and/or periodontitis occur in some patients/clients who use inhaled corticosteroids for long periods of time or at high dose. The use of steroid inhalers can also result in throat irritation, voice impairment, cough, dry mouth, and, rarely, tongue enlargement. Patients/clients using inhaled medications are also at increased risk of dental erosion and periodontal disease.

■ Pancreatic enzyme replacement treatment can lead to irritation of the oral mucosa if the capsule contents are (inappropriately) held in the mouth.

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1 Coryza (or rhinitis) is irritation and inflammation of the mucous membranes of the nose. Common signs/symptoms are nasal congestion, runny nose, sneezing, and post-nasal drip.

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Related signs and symptoms

- Cystic fibrosis is an autosomal recessive disorder that primarily affects Caucasians of Northern and Western European ancestry. It is the most common lethal genetic disease affecting Canadian children and young adults, with the median age of survival being about 49 years of age.

- About 4,000 Canadians live with CF, and more than 100 children are newly diagnosed annually in Canada (about 1 in every 3,600 births). While disease prevalence is holding steady, the median age is shifting upward given the advent of genetic testing, prenatal testing, earlier detection via newborn screening, and better treatments. Nearly 60% of all persons with CF in Canada are adults.

- The disease affects all exocrine (secretory) glands, and is characterized by the production of thick, sticky mucus that causes blockages in the lungs, pancreatic ducts, and other organs. This situation leads to an environment that facilitates the overgrowth of bacteria in affected tissues, particularly in the lungs and sinuses.

- Cystic fibrosis is a multi-system disorder that produces a variety of signs and symptoms including:
  - failure to thrive in infancy and childhood;
  - persistent cough with productive thick mucus;
  - wheezing and shortness of breath;
  - frequent respiratory tract infections, including pneumonia, bronchitis, and bronchiectasis;
  - bowel disorders, such as intestinal obstruction or frequent, foul-smelling, oily, loose stools;
  - weight loss or failure to gain weight despite possible increased appetite;
  - salty tasting sweat; and
  - infertility (males) and decreased fertility (females).

- Episodic intestinal blockage is frequent in newborns and young children.

- Complications and sequelae of CF in adolescence and adulthood include: CF-related diabetes (CFRD); liver disease (sometimes requiring liver transplantation); distal intestinal obstruction syndrome (i.e., build-up of stool in the intestinal tract secondary to mucus plugging); osteoporosis (associated with poor vitamin D absorption, poor nutritional status, physical inactivity, and corticosteroid therapy); nasal polyps (resulting in bilateral nasal blockage, runny nose, decreased sense of smell, facial pain, congestion, and, less frequently, nose bleeds); gallstones; pancreatitis; rectal prolapse (caused by frequent coughing and defecation problems); pneumothorax; and clubbing of the fingers (which is indicative of continual poor oxygenation of the blood). Depression and anxiety are also common.

- The four main approaches to CF treatment are nutritional repletion (which includes pancreatic enzyme replacement therapy, fat soluble vitamin supplementation, and high-fat, high-carbohydrate diet); clearance of airway obstruction (via chest physical therapy, exercise, and bronchodilator medications); addressing airway infection (via ad hoc antibiotic treatment); and suppressing infection (via long-term antibiotic prophylaxis).

- For some persons with advanced CF, organ transplantation may be an option. Currently, double-lung, heart-lung and liver transplants are the only definitive treatments for CF patients/clients with progressive disease.

- All newborns in Ontario are specifically screened for cystic fibrosis via their newborn blood-screening test. As well, if a physician suspects a patient/client has CF, a “sweat test” (to detect elevated salt content) may be administered, and a test for the presence of enzymes in the intestine can be performed. Without early detection, irreversible damage to the lungs and digestive system may occur.

- There are 15 specialized CF clinics (pediatric and adult) in Ontario, and an additional 27 nation-wide.

- Respiratory failure is the most common cause of death.

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2 Bronchiectasis is characterized by enlargement and “pocketing” of the bronchi/bronchial tubes, in which mucus collects and bacteria breed, thereby leading to recurrent lung infections and cumulative damage to the airways.
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References and sources of more detailed information

- Cystic Fibrosis Canada [www.cysticfibrosis.ca](http://www.cysticfibrosis.ca)

* Includes oral hygiene instruction, fitting a mouth guard, taking an impression, etc.
** Ontario Regulation 501/07 made under the Dental Hygiene Act, 1991. Invasive dental hygiene procedures are scaling teeth and root planing, including curetting surrounding tissue.

Date: November 3, 2015