Use of the dental hygiene interventions of scaling of teeth and root planing including curetting surrounding tissue, orthodontic and restorative practices, and other invasive interventions for persons\(^1\) with cystic fibrosis.

**ADVISORY OBJECTIVE(S)**

To guide dental hygienists at the point of care relative to the use of the Procedures for persons who have cystic fibrosis, chiefly as follows.

1. Understanding the medical condition.
2. Sourcing medications information.
3. Taking the medical and medications history.
4. Identifying and contacting the most appropriate healthcare provider(s) for medical advice.

\(^1\) Persons includes young persons and children
5. Understanding and taking appropriate precautions prior to and during the Procedures proposed.
6. Deciding when and when not to proceed with the Procedures proposed.
7. Dealing with adverse events arising during the Procedures.
8. Record keeping.

TARGET POPULATION

Child (2 to 12 years)
Adolescent (13 to 18 years)
Adult (19 to 44 years)
Middle Age (45 to 64 years)
Aged (65 to 79 years)
Aged, 80 and over
Male
Female
Parents, guardians, and family caregivers of children, young persons and adults with cystic fibrosis.

MAJOR OUTCOMES CONSIDERED

For persons who have cystic fibrosis: to maximize health benefits and minimize adverse effects by promoting the performance of the Procedures at the right time with the appropriate precautions, and by discouraging the performance of the Procedures at the wrong time or in the absence of appropriate precautions.

RECOMMENDATIONS

UNDERSTANDING THE MEDICAL CONDITION

Terminology used in this Advisory

Resources consulted
- Cystic Fibrosis Canada
- US National Library of Medicine and the National Institutes of Health Medline Plus

1. Cystic fibrosis is
   a. a multi-organ disease of the mucus and sweat glands that chiefly affects the lungs, pancreas, liver, intestines, sinuses and sex organs
   b. the most common fatal genetic disease affecting young Canadians
   c. also termed
      i. CF
      ii. Cystic fibrosis of the pancreas
      iii. Fibrocystic disease of the pancreas
      iv. Mucoviscidosis
      v. Mucoviscidosis of the pancreas
      vi. Pancreas fibrocystic disease
      vii. Pancreatic cystic fibrosis
2. Bronchiectasis, pockets in the bronchial tubes where mucus collects and in which bacteria breed, from which originate repeated lung infections with cumulating damage to the bronchial tubes; untreated, bronchiectasis may lead to respiratory failure or other serious illnesses.

3. Palliative care, services of care for persons towards the end of life with terminal illnesses such as cancer, when the focus of the care
   a. is relieving symptoms
   b. attending to physical and spiritual needs.

4. Pneumothorax, collapsed lung.

5. Supportive care, services of care to help persons meet the physical, emotional and spiritual challenges arising from the condition or its treatment.

Overview of cystic fibrosis

Resources consulted

- Canadian Cystic Fibrosis Foundation
- US National Library of Medicine and the National Institutes of Health Medline Plus

Cystic fibrosis

1. Affects both males and females.
2. Affects persons from all racial and ethnic groups but is most common among Caucasians whose ancestors came from northern Europe.
3. Has no cure but its treatments have improved since 1980 raising life expectancy from childhood, teenage or young adulthood to a median age of survival of 37 for Canadians, as at 2002.
4. Varies widely in symptoms and severity; with the milder versions, the diagnosis may not be made until teenage or young adulthood; is nevertheless diagnosed by the age of 10 in 90 percent of patients/clients.
5. Affects an estimated 1 in 3,600 children born in Canada.
6. Requires nearly 4,000 children, adolescents, and adults with cystic fibrosis to attend specialized clinics in Canada.
7. Results in respiratory failure as its most common immediate cause of death.
8. Causes severe breathing problems associated with build-up of thick mucus that
   a. impedes clearance of bacteria and leads to cycles of infection and inflammation
   b. leads to damage of lung tissues
   c. requires of persons with the disease a demanding daily routine of physical therapy to keep the lungs free of congestion and infection.
9. Seriously impairs digestion and absorption of adequate nutrients and vitamins from food because thick mucus blocks the ducts of the pancreas
   a. preventing enzymes from reaching the intestines to digest food
   b. requiring persons with the disease to take by mouth numerous artificial enzymes with every meal and snack.
10. Is manifested as
    a. difficulty breathing
    b. constant cough productive of thick mucus
    c. excessive appetite, with poor weight gain or even weight loss
    d. persistent diarrhea or bulky, foul-smelling, and greasy stools
    e. intestinal gas, abdominal swelling, and pain or discomfort
f. excessively salty sweat, causing mineral imbalance  
g. repeated or prolonged bouts of pneumonia  
h. failure to thrive.  

11. In diagnosis, may be confused with  
a. Asthma (CDHO Advisory)  
b. chronic bronchitis  
c. pneumonia  
d. celiac disease (CDHO Advisory).  

12. Is caused by a defect in a gene carried by 1 in 25 Canadians, termed carriers, who do not have the disease  
a. when both parents are carriers, there is a  
   i. 25 percent chance that their child will be born with cystic fibrosis  
   ii. 50 percent chance that their child will not have cystic fibrosis, but will be a carrier  
   iii. 25 percent chance that their child will not have cystic fibrosis, and will not be a carrier  
b. with each pregnancy, the risks are identical; carrier parents may have several children with cystic fibrosis, or none.  

13. Is treated at home with  
a. vigorous percussion of the chest and the back  
b. positive expiratory pressure mask therapy or other forms of chest physiotherapy to help loosen mucus  
c. pancreatic enzymes with all meals  
d. nutritional supplements and vitamins  
e. antibiotics in oral, intravenous and or inhaled forms, to ease congestion and for prophylaxis  
f. exercise.  

14. Has no known cure but, since 1989 when Canadian researchers discovered the gene responsible, global research to find a cure for the disease brings optimism to the efforts to find a solution.  

Comorbidity, complications and associated conditions  

Comorbid conditions are those which co-exist with cystic fibrosis but which are not believed to be caused by it. Complications and associated conditions are those that may have some link with it. Distinguishing among comorbid conditions, complications and associated conditions may be difficult in clinical practice.  

Comorbidities, complications and conditions associated with cystic fibrosis include  
1. Sinusitis: arises from swelling in and blockage of the sinuses; most people with cystic fibrosis develop sinusitis  
2. Bronchiectasis  
3. Nasal polyps, often requiring surgery  
4. Finger clubbing, indicative of inadequate oxygenation of the blood stream.  
5. Pneumothorax  
6. Pancreatitis  
7. Diabetes (CDHO Advisory)  
8. Gallstones
9. Liver disease secondary to inflammation or blockage of bile ducts (*CDHO Advisory*).
10. Episodic intestinal blockage, especially in newborns.
11. Rectal prolapse caused by frequent coughing or problems with defecation.
12. Low bone mineral density associated with low Vitamin D intake (*CDHO Advisory*).

**Oral health considerations**

**Resources consulted**

- Oral health and related factors in cystic fibrosis and other chronic respiratory disorders
- Pathogenesis of Early Lung Disease in Cystic Fibrosis: A Window of Opportunity To Eradicate Bacteria

1. A partially controlled study of the oral conditions of 42 patients/clients in treatment for cystic fibrosis observed, among other things, that
   a. compared to the control group, in which every person had some amount of plaque or gingival disease, the patient/client group had a lowered incidence of plaque and less gingival disease
   b. minimal plaque in the patient/client group could be related to factors such as the life-long use of various antibiotic agents, the chewing of digestive enzyme supplements, the effect of medical management on tooth hardness, and the effect of stained teeth on the plaque microorganisms
   c. the therapy for cystic fibrosis was apparently beneficial to the periodontal health of the patients/clients group.

2. Other research
   a. points to biofilms, a defensive mode of bacterial growth which resists treatment and eradication of bacterial communities
   b. explains that biofilms
     i. are communities of bacteria enclosed in a self-produced matrix attached to a surface
     ii. include dental plaque, endocarditis, and slime on river stones
     iii. are increasingly recognized as contributing to the pathogenesis of cystic fibrosis and as having a role in other bacterial diseases
     iv. protect bacteria in that they exhibit increased resistance to antibiotics and host defense factors
     v. may prevent clinically attainable antibiotic concentrations from adequately clearing biofilm infections, allowing the bacterial population to recover, persist, and spread.

**MEDICATIONS SUMMARY**

**Sourcing medications information**

1. Adverse effect database
   - Health Canada’s Marketed Health Products Directorate
toll-free 1-866-234-2345
   - Health Canada’s Drug Product Database
2. Specialized organizations
   - US National Library of Medicine and the National Institutes of Health Medline Plus Drug Information
   - WebMD

3. Medications considerations
   All medications have potential side effects whether taken alone or in combination with other prescription medications, or as over-the-counter (OTC) or herbal medications.

4. Information on herbals and supplements
   - US National Library of Medicine and the National Institutes of Health Medline Plus Drug Information All Herbs and Supplements

Types of medications

Warnings
Individual medications may be subject to important warnings, which
1. change from time to time
2. may affect the appropriateness, efficacy or safety of the Procedures
3. are accessible via the links to the particular medications listed below or through the specialized organizations listed above
4. through the links, should be viewed by dental hygienists in the course of their familiarizing themselves about a medication or combination of medications identified in the patient/client’s medical and medications history.

Overall aims of medication therapy for cystic fibrosis
1. help keep the lungs as healthy as possible
2. reduce and control mucus in the lungs
3. replace digestive enzymes.

Medication therapy includes
1. Treating infections with antibiotics such as
   - azithromycin (Zithromax®)
   - cephalexin (Keflex® Pulvules®)
   - ciprofloxacin (Cipro®, Cipro® XR, Proquin® XR)
   - tobramycin (Nebcin®, Tobi®).
2. Opening and keeping open airways with
   a. bronchodilators such as
      - salbutamol, albuterol (VoSpire ER®)
      - albuterol inhalation (Ventolin HFA®, Proventil® HFA)
      - salmeterol (Serevent®)
   b. anticholinergics such as
      - ipratropium (Atrovent®).
3. Controlling the amount and thickness of mucus with medications such as
   - dornase alfa (Pulmozyme®).
4. Reducing inflammation with
   a. non-steroidal anti-inflammatory drugs (NSAIDs), see usage note, such as ibuprofen
   b. membrane stabilizers such as
      - cromolyn sodium oral inhalation (Intal®)
nedocromil (Tilade® Inhaler)

c. corticosteroids, see usage note for oral and inhaled, such as
   prednisone
   methylprednisolone oral (Medrol®, Meprolone®)
   fluticasone oral inhalation (Flovent® HFA).

5. Replacing the effect of digestive enzymes with enzyme replacement therapy such as pancrelipase (for example, Creon® or Pancrease®).

6. Helping (in persons with G551D mutation of CF) defective protein CFTR (cystic fibrosis transmembrane conductance regulator) work at the surface of the cells to decrease build-up of thick mucus, such as ivacaftor (Kalydeco™)

Side effects of medications

See also the links to the specific medications above

1. Oral corticosteroids are
   a. recommended only for people who have significant shortness of breath and wheezing, or the fungus infection, allergic bronchopulmonary aspergillosis
   b. not recommended for long-term use because of potentially serious side effects, such as
      i. growth retardation in children
      ii. diabetes (CDHO Advisory)
      iii. cataracts
      iv. osteoporosis (CDHO Advisory)
      v. decreased ability to fight lung infections.

2. Inhaled steroids are recommended chiefly for persons with cystic fibrosis and asthma.

3. Non-steroidal anti-inflammatory drugs (NSAIDS) are
   a. recommended chiefly for persons with cystic fibrosis because the side effects are fewer and less serious than those of steroids used as anti-inflammatory medications, though with long-term use they may be responsible for internal bleeding or kidney problems
   b. available as Ibuprofen, over the counter and on prescription, which is believed in long-term use to slow the loss of lung function in cystic fibrosis; for this reason it is recommended only in prescription form after consultation with the prescribing physician.

THE MEDICAL AND MEDICATIONS HISTORY

The medical and medications history-taking should

1. Focus on screening the patient/client prior to treatment decision relative to
   a. key symptoms, and severity
   b. medications considerations
   c. contraindications
   d. complications
   e. comorbidities.

2. Explore the need for advice from the appropriate primary or specialized care
provider(s).
3. Inquire about
   a. antibiotic prophylaxis
   b. the patient/client’s understanding and acceptance of the need for oral healthcare
   c. medications considerations, including over-the-counter medications, herbals and supplements
   d. problems with previous dental/dental hygiene care
   e. problems with infections generally and specifically associated with dental/dental hygiene care
   f. the patient/client’s current state of health
   g. how the patient/client’s current symptoms relate to
      i. oral health
      ii. health generally
      iii. recent changes in the patient/client’s condition.

IDENTIFYING AND CONTACTING THE MOST APPROPRIATE HEALTHCARE PROVIDER(S) FOR ADVICE

Identifying and contacting the most appropriate healthcare provider(s) from whom to obtain medical or other advice pertinent to a particular patient/client

   1. Record the name of the physician/primary care provider most closely associated with the patient/client’s healthcare, and the telephone number.
   2. Obtain from the patient/client or parent/guardian written, informed consent to contact the identified physician/primary healthcare provider.
   3. Use a consent/medical consultation form, and be prepared to fax the form to the provider.
   4. Include on the form a standardized statement of the Procedures proposed, with a request for advice on proceeding or not at the particular time, and any precautions to be observed.

UNDERSTANDING AND TAKING APPROPRIATE PRECAUTIONS

Infection control

Dental hygienists are required to keep their practices current with infection control policies and procedures, especially in relation to

   1. The Recommendations published by the Centers for Disease Control and Prevention (a frequently updated resource).
   2. Relevant occupational health and safety legislative requirements.
   3. Relevant public health legislative requirements.
   4. Best practices or other protocols specific to the medical condition of the patient/client.

DECIDING WHEN AND WHEN NOT TO INITIATE THE PROCEDURES PROPOSED

   1. Antibiotic prophylaxis may be required (particularly for patients/clients who are on immunosuppressants for heart, lung, and liver transplants) so the dental hygienist should consult with the primary care physician to obtain advice about implementing the Procedures.
   2. For a patient/client whose symptoms are under control and who does not require
antibiotic prophylaxis, the dental hygienist should implement the Procedures, though these may be postponed pending medical advice, which is likely to be required
   a. symptoms or signs of exacerbation of the medical condition
   b. comorbidity, complication or an associated condition of cystic fibrosis
   c. not recently or ever sought and received medical advice relative to oral healthcare procedures
   d. recently changed significant medications, under medical advice or otherwise
   e. recently experienced changes in his/her medical condition such as medication or other side effects of treatment
   f. is deeply concerned about any aspect of his or her medical condition.

DEALING WITH ANY ADVERSE EVENTS ARISING DURING THE PROCEDURES

Dental hygienists are required to initiate emergency protocols as required by the College of Dental Hygienists of Ontario’s Standards of Practice, and as appropriate for the condition of the patient/client.

First-aid provisions and responses as required for current certification in first aid.

RECORD KEEPING

Subject to Ontario Regulation 9/08 Part III.1, Records, in particular S 12.1 (1) and (2)

For a patient/client with a history of cystic fibrosis, the dental hygienist should specifically record
1. A summary of the medical and medications history.
2. Any advice received from the physician/primary care provider relative to the patient/client’s condition.
3. The decision made by the dental hygienist, with reasons.
4. Compliance with the precautions required.
5. All Procedure(s) used.
6. Any advice given to the patient/client.

ADVISING THE PATIENT/CLIENT

The patient/client is urged to alert any healthcare professional who proposes any intervention or test that he or she has a history of cystic fibrosis.

As appropriate, discuss
1. The importance of the patient/client’s
   a. self-checking the mouth regularly for suspicious signs or symptoms
   b. reporting to the appropriate healthcare provider any changes in the mouth indicative of suspicious lesions.
2. The importance of infection control within the oral cavity.
3. The need for regular oral health examinations and preventive oral healthcare.
4. Oral self-care including information about
   a. choice of toothpaste
   b. tooth-brushing techniques and related devices
   c. dental flossing
   d. mouth rinses
   e. management of a dry mouth.
5. The importance of an appropriate diet in the maintenance of oral health.
6. For persons at an advanced stage of a disease or debilitation
   a. regimens for oral hygiene as a component of support care and palliative care
   b. the role of the family caregiver, with emphasis on maintaining an infection-free environment through hand-washing and, if appropriate, wearing gloves
   c. scheduling and duration of appointments to minimize stress and fatigue.
7. Comfort level while reclining, and stress and anxiety related to the Procedures.
8. Medication side effects such as dry mouth, and recommend treatment.
9. Mouth ulcers and other conditions of the mouth relating to cystic fibrosis, comorbidities, complications or associated conditions, medications or diet.

### BENEFITS/HARMS OF IMPLEMENTING THE RECOMMENDATIONS

#### POTENTIAL BENEFITS

1. Promotion of health through oral hygiene for persons who have cystic fibrosis.
2. Reduction of the adverse effects, such as exacerbation of breathing difficulties by
   a. paying particular attention to infection control
   b. generally increasing the comfort level of persons in the course of dental hygiene interventions
   c. using appropriate techniques of communication
   d. providing advice on scheduling and duration of appointments.
3. Reduction of risk of oral health needs being unmet.

#### POTENTIAL HARMs

1. Causing infection in the respiratory system of persons with cystic fibrosis.
2. Performing the Procedures at an inappropriate time, such as
   a. during a period of exacerbated breathing difficulty
   b. in the presence of complications for which prior medical advice is required
   c. in the presence of acute oral infection without prior medical advice.
3. Disturbing the normal dietary and medications routine of a person with cystic fibrosis.
4. Inappropriate management of pain or medication.

### CONTRAINDICATIONS

#### CONTRAINDICATIONS IN REGULATIONS

Identified in the Dental Hygiene Act, 1991 – O. Reg. 218/94 Part III

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