### Advisory Title

Use of the dental hygiene interventions of scaling of teeth and root planing including curettage of surrounding tissue, orthodontic and restorative practices, and other invasive interventions for persons with sickle cell disease.

### Advisory Status

Cite as:
*College of Dental Hygienists of Ontario, CDHO Advisory Sickle Cell Disease, 2010-07-15*

### Interventions and Practices Considered

Scaling of teeth and root planing including curettage of surrounding tissue, orthodontic and restorative practices, and other invasive interventions (“the Procedures”).

### Scope

**Disease/Condition(s)/Procedure(s)**

*Sickle cell disease*

### Intended Users

- Advanced practice nurses
- Dental assistants
- Dental hygienists
- Dentists
- Denturists
- Dieticians
- Health professional students
- Nurses
- Patients/clients
- Pharmacists
- Physicians
- Public health departments
- Regulatory bodies

### Advisory Objective(s)

To guide dental hygienists at the point of care relative to the use of the Procedures for persons who have sickle cell disease, 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.

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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)
Male
Female

Parents, guardians, and family caregivers of children, young persons and adults with sickle cell disease.

MAJOR OUTCOMES CONSIDERED

For persons who have sickle cell disease: 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

- Government of Ontario: Hemoglobinopathies, Sickle Cell Disease (HbSS, HbSC or HbS/ß-Thalassemia)
- HealthLinkBC: Sickle Cell Disease
- NIH: Genetics Home Reference, Sickle cell disease
- Sickle Cell Disease Association of America: What is Sickle Cell Disease?
- Suite 101: Sickle Cell Disease, Features and Management of a Genetic Red Blood Cell Disorder
- The Sickle Cell Association of Ontario: What Is Sickle Cell Anemia?
- University of Illinois College of Medicine: What is Sickle Cell Disease?

Terminology varies among centres; that used here is selected for the purposes of this Advisory.

1. Sickle cell disease, a group of inherited blood disorders, associated with the inheritance of two sickle cell genes, one from each parent, characterized by
   a. atypical hemoglobin molecules
   b. signs and symptoms that
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i. usually begin in early childhood, including
   1. reduction in circulating red blood cells
   2. repeated infections
ii. vary in severity of symptoms from person to person ranging from mild to severe
   c. sickling of red blood cells, which causes them to break down prematurely, which may
      i. lead to anemia (CDHO Advisory) causing
         1. shortness of breath
         2. fatigue
         3. delayed growth and development in children
      ii. cause yellowing of the eyes and skin, the signs of jaundice
   d. painful episodes that
      i. occur when sickled red blood cells, which are stiff and inflexible, lodge in and block small blood vessels
      ii. are cyclical in nature
      iii. result from deprivation of oxygen in tissues and organs, leading to organ damage, especially in the lungs, kidneys, spleen, and brain
   e. particularly serious complications in the lungs
      i. high blood pressure in the blood vessels that supply the lungs, which
         1. occurs in about one-third of adults with sickle cell disease
         2. can lead to heart failure
      ii. acute chest syndrome, sudden blockage of a blood vessel in the lungs that
         1. is associated with fever or respiratory symptoms
         2. affects about 40 percent persons with sickle cell disease
         3. is most common reason for early death.

Other terminology
1. Acute sickle cell pain episode, sickle cell crisis, pain lasting from several minutes to several days, caused by blood flow blocked with sickled red blood cells.
2. Chronic pain from sickle cell bone damage
   a. lasts longer than a few weeks, possibly present every day
   b. occurs when bones are damaged by blocked blood flow.
3. Chronic nerve pain in sickle cell disease
   a. arises from damage to the nerves from blocked blood flow
   b. creates burning, tingling, and numbing discomfort, possibly present every day.
4. Gene mutations related to sickle cell disease
   a. in one particular gene, mutations create abnormal versions of hemoglobin that distort red blood cells into a sickle shape, which causes them to
      i. die prematurely, which can lead to anemia (CDHO Advisory)
      ii. lodge in and block small blood vessels causing serious medical complications
   b. one particular mutation produces abnormal hemoglobin known as hemoglobin S (HbS)
   c. other mutations lead to
      i. abnormal versions of hemoglobin such as
         1. hemoglobin C (HbC)
         2. hemoglobin E (HbE)
ii. abnormally low level of hemoglobin, a condition called beta thalassemia.

5. Hand-foot syndrome, swelling of hands and feet
   a. developed by almost all individuals with sickle cell disease
   b. most common in childhood
   c. arises from poor blood flow, and may lead to loss of tips of fingers and toes.

6. HbS disease, alternate name for sickle cell disease.

7. Hemoglobin S Disease, alternate name for sickle cell disease.

8. Immunosuppression (CDHO Advisory), suppression of immunity with medications, its main drawback is the increased risk of infection for the duration of treatment.

9. Lesion, a term variously and loosely used in medicine to refer to such things as
   a. any abnormality of tissue in the body, including the mouth and skin
   b. any localized abnormal structural change in a bodily part
   c. a mass especially before a definite diagnosis is established
   d. cancer
   e. an injury to living tissue, such as a cut or break in the skin.

10. Mucositis is
    a. inflammation of the mucous membranes lining the digestive tract from the mouth to the anus
    b. a common side effect of chemotherapy (CDHO Advisory) and of radiation therapy (CDHO Advisory) that may involve the entire gastrointestinal tract, leading to one or more of
       i. painful mouth sores
       ii. diarrhea
       iii. nausea
       iv. abdominal pain in any part of the digestive tract
    c. one of the most common adverse effects of transplantation.

11. Oral ulcer, an open lesion, often painful, inside the mouth or upper throat, an alternative name for
    a. a mouth ulcer
    b. an aphthous ulcer
    c. aphthous stomatitis, also known as a canker sore
    d. a cancerous ulcer.

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

13. Risk factor, a term
    a. used strictly to identify anything that affects the person’s chances of developing a disease
    b. used loosely to refer to things that in themselves may not affect the person’s chances of developing a disease but which may signal the presence of things that are risk factors as strictly defined
    c. for information used for medical history-taking and examination
    d. that, in the absence of clear-cut epidemiological data, often cannot be quantified or even rank-ordered for importance.

14. Sickle cell anemia, alternate name for sickle cell disease.

15. Sickle cell disorders, alternate name for sickle cell disease.
16. Sickle cell trait, where the person, the carrier, inherits only one gene, is asymptomatic.
17. Sickling, the distortion of red blood cells into a sickle, or crescent, shape.
18. Sickling disorder due to hemoglobin S, alternate name for sickle cell disease.
19. Supportive care, services of care to help persons meet the physical, emotional and spiritual challenges arising from the condition or its treatment.

Overview of sickle cell disease

Resources consulted
- The Hospital for Sick Children: Sickle Cell Anemia
- US NIH National Heart, Lung, and Blood Institute: Sickle Cell Anemia
- National Institutes of Health: The Management of Sickle Cell Disease, Fourth Edition

Sickle cell disease

1. Affects millions of people worldwide; is most common among people whose ancestors come from
   a. Africa
   b. Mediterranean countries such as Greece, Turkey, and Italy
   c. The Arabian Peninsula
   d. India
   e. Spanish-speaking regions in South America, Central America, and parts of the Caribbean.

2. Is the most common inherited blood disorder in the United States, which
   a. affects males and females equally
   b. affects 70,000 to 80,000 Americans
   c. is estimated to occur in 1 in 500 African Americans and 1 in 1,000 to 1,400 Hispanic Americans.

3. Commonly presents
   a. first signs and symptoms in early childhood, as early as 4–9 months
   b. as anemia (CDHO Advisory) and pain
   c. as sudden pain throughout the body, a sickle cell crisis, often affecting the bones, lungs, abdomen, and joints.

4. Lacks a widely available cure
   a. symptomatic treatments include medications, fluids, and pain relief
   b. bone marrow transplants may offer a cure in a small number of cases.

5. Cannot be prevented because it is an inherited disease, but steps can be taken to reduce its complications.

6. With good health care, is compatible with reasonably good health and productive lives which last longer than in the past; but, despite all the innovations in treatment, the majority of affected persons do not survive beyond the age of 50.

7. Is a common reason for individuals to seek emergency medical care, presents in varied forms, and creates a diagnostic dilemma for the unwary emergency room physician.

8. Varies widely in its precipitating factors for crises, making these unpredictable and variable in presentation: factors include
   a. stress
   b. extremes of weather
   c. infection
9. Should be supported by a healthy lifestyle, steps to prevent and control complications, and techniques for coping with pain.

10. Is diagnosed with a blood test for hemoglobin S, and is confirmed by microscopy to identify the sickle cells.

11. Is treated with emphasis on prevention of the sickle cell crises and their symptoms and complications, with
   a. oxygen
   b. fluids
   c. pain control
   d. antibiotics
   e. blood transfusions
   f. medications.

12. Requires parents to learn about the disease and help the child manage it.

Comorbidity, complications and associated conditions

Comorbid conditions are those which co-exist with sickle cell disease 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 because any major organ may be affected by sickle cell disease.

1. Liver, heart, kidneys, gallstone, eyes, bones, and joints may suffer damage from the abnormal function of the sickle cells and their inability to flow smoothly through the small blood vessels.

2. Comorbidities, complications and potential side-effects of treatment include
   a. acute chest syndrome
   b. anemia (CDHO Advisory)
   c. bone damage
   d. eye damage
   e. frequent infections
   f. gallstones (CDHO Advisory)
   g. jaundice (CDHO Advisory)
   h. kidney damage and loss of body water in the urine (CDHO Advisory)
   i. leg ulcers
   j. pain
   k. stroke (CDHO Advisory)
   l. stunted growth.

Oral health considerations

Resources consulted

- BlackHealthCare.com: Sickle Cell Anemia - Case Management
- Journal of Indian Society of Pedodontics and Preventive Dentistry: Dental considerations in the management of children suffering from sickle cell disease, A case report
1. Routine dental care and oral hygiene are important to prevent loss of teeth and infections that may lead to further complications of sickle cell disease.
2. Dental caries and other problems are associated with sickle cell disease.
3. Dental procedures that require local anesthesia can be performed in the dental office as with any other patient/client.
4. Patients/clients with a history of heart disease may require antibiotic prophylaxis prior to the Procedures.
5. Stress should be minimized because this may provoke a sickle cell crisis.
6. Acute conditions associated with sickle cell disease
   a. are generally of such severity that they are unlikely to be encountered in normal dental hygiene practice
   b. should always be viewed as exhibiting a contraindication to the Procedures
   c. require that anyone, children included, presenting for dental hygiene care with a fever, defined as a body temperature above 38° C, and a history of sickle cell disease should be
      i. viewed as unsuitable for dental hygiene, and
      ii. referred as an emergency to a hospital.
7. Complications, comorbidities and side effects associated with sickle cell disease
   a. are increasingly likely to be encountered in dental hygiene practice as
      i. life expectancy grows with improvements in healthcare
      ii. awareness increases in the importance of oral hygiene in sickle cell disease and generally
   b. require consideration of the risks of provoking sickle cell crises
   c. require exploration of the need for antibiotic prophylaxis for the Procedures.
8. Bone marrow transplant is a contraindication to the Procedures because it variously involves
   a. chemotherapy (CDHO Advisory) or radiation therapy (CDHO Advisory) to destroy the host’s bone marrow
   b. life-long need for immunosuppression to prevent rejection
   c. oral side effects such as mucositis.

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

5. Complementary and alternative medicine
   - National Center for Complementary and Alternative Medicine

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.

Medications
   1. Pain medications for sickle cell crises, acute sickle cell pain episode
      a. opioid medications
         i. mild opiates
            - **codeine** (Fiorinal® with Codeine, as a combination product containing codeine phosphate, aspirin, butabarbital, and caffeine, among others)
            *blocks fever and may therefore conceal an emergency
            **acetaminophen** combined with codeine (Tylenol with Codeine (No. 2, No. 3, No. 4)®, among others)
            *blocks fever and may therefore conceal an emergency
            **hydrocodone** (Vicodin® containing hydrocodone and acetaminophen, among others)
            *blocks fever and may therefore conceal an emergency
      b. non-steroid anti-inflammatory drugs
         - aspirin (ASA, acetylsalicylic acid)
         *blocks fever and may therefore conceal an emergency
         **ibuprofen** (Advil®, Motrin®)
         *blocks fever and may therefore conceal an emergency
      c. anti-neoplastic agents that
         i. reduce the frequency of pain crises and acute chest syndrome
ii. decrease the need for frequent blood transfusions  
iii. include hydroxyurea (Droxia®, Hydrea®)  

2. Pain medications for chronic pain from sickle cell bone damage  
   a. long-acting arthritis medications, non steroidal anti-inflammatory drugs  
      celecoxib (Celebrex®)  
      ibuprofen (Advil®, Motrin®)  
      salsalate (disalicylic acid, salicylsalicylic acid)  
   b. long-acting opiates, narcotic analgesics, where NSAIDS are insufficient  
      methadone (Dolophine®, Methadose®)  
      morphine (Avinza®, MS Contin®)  
      oxycodone (OxyContin®, Roxicodone®, among others)  

3. Tricyclic antidepressants, such as  
   amitriptyline (Limbitrol®)  

4. Corticosteroids  
   prednisone (Prednisone Intensol®, Sterapred®)  

5. Anticonvulsants, such as  
   gabapentin (Gabarone®, Neurontin®)  

6. Antibiotics, such as  
   penicillin V potassium oral (Beepen-VK®, Betapen-VK®, among others)  

7. Vitamins, such as  
   folic acid (Folvite®)  

Side effects of medications  
See the links above to the individual medications.  

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  
   b. medications  
   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. symptoms indicative of raised body temperature  
   b. symptoms indicative of impending a sickle cell crisis  
   c. requirements for antibiotic prophylaxis  
   d. past, current or impending treatment with immunosuppression  
   e. the patient/client’s experience with emergencies associated with sickle cell disease.  
   f. the patient/client’s understanding and acceptance of the need for oral healthcare  
   g. medications considerations, including over-the-counter medications, herbals and supplements
h. problems with previous dental/dental hygiene care
i. problems with infections generally and specifically associated with dental/dental hygiene care
j. the patient/client’s current state of health
k. 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.
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

The dental hygienist
1. should not implement the Procedures
   a. without prior consultation with the appropriate primary or specialist care provider(s) because sickle cell disease is a blood disorder
   b. if the patient/client has a body temperature above 38° C, which constitutes a medical emergency requiring
      i. a 911 call
      ii. referral to a hospital emergency department accompanied by a brief referral note that
         1. identifies the patient/client as having a history of sickle cell disease
         2. mentions the raised body temperature
c. if the patient/client has undergone or is shortly to undergo bone marrow transplantation
d. if the patient/client requires antibiotic prophylaxis.

2. may postpone the Procedures pending medical advice if the patient/client
   a. appears debilitated
   b. reports complications of sickle cell disease
   c. has not complied with pre-medication, including antibiotic prophylaxis, as directed by the prescribing physician
d. has 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 unable to provide the dental hygienist with sufficient information about
   i. medications
   ii. treatment
g. has symptoms or signs of
   i. exacerbation of the medical condition
   ii. comorbidity, complication or an associated condition of liver disease
h. not recently or ever sought and received medical advice relative to oral healthcare procedures
   i. is deeply concerned about any aspect of his or her medical condition.

DEALING WITH ANY ADVERSE EVENTS ARISING DURING THE PROCEDURES

Specific to sickle cell disease

Sudden pain throughout the body may signal the onset of a sickle cell crisis the patient/client should be
1. transported by ambulance, 911 call, to the Emergency Room of a hospital
2. accompanied by a brief referral note that
   a. identifies the patient/client as having a history of sickle cell disease
   b. mentions the sudden onset of pain.

General

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 sickle cell disease, 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 sickle cell disease.

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 need for regular oral health examinations and preventive oral healthcare especially for children.
3. 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.
4. The importance of an appropriate diet in the maintenance of oral health.
5. 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.
6. Comfort level while reclining, and stress and anxiety related to the Procedures.
7. Medication side effects such as dry mouth, and recommend treatment.
8. Mouth ulcers and other conditions of the mouth relating to sickle cell disease, comorbidities, complications or associated conditions, medications or diet.

BENEFITS/HARMS OF IMPLEMENTING THE RECOMMENDATIONS

POTENTIAL BENEFITS

1. Promoting health through oral hygiene for persons who have sickle cell disease.
2. Reducing the adverse effects, such as prevention of stress and thus a potential sickle cell crisis by
   a. generally increasing the comfort level of persons in the course of dental hygiene interventions
   b. using appropriate techniques of communication
   c. providing advice on scheduling and duration of appointments.
3. Reducing the risk that oral healthcare needs are unmet.
### POTENTIAL HARMs

1. Causing harm through failure to recognize that the patient/client has a body temperature above 38°C.
2. Performing the Procedures at an inappropriate time, such as
   a. when a sickle cell crisis is impending
   b. when the patient/client is at increased risk of infection
   c. in the presence of comorbidities and complications for which prior medical advice is required
   d. in the presence of acute oral infection without prior medical advice.
3. Disturbing the normal dietary and medications routine of a person with sickle cell disease.
4. Inappropriate management of pain or medication.

### CONTRAINDICATIONS

Identified in the [Dental Hygiene Act, 1991 – O. Reg. 218/94 Part III](#)

### ORIGINALLY DEVELOPED

2009-11-24

### DATE OF LAST REVIEW

2010-07-15

### ADVISORY DEVELOPER(S)

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### SOURCE(S) OF FUNDING

College of Dental Hygienists of Ontario

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# ACKNOWLEDGEMENTS

The College of Dental Hygienists of Ontario gratefully acknowledges the *Template of Guideline Attributes*, on which this advisory is modelled, of *The National Guideline Clearinghouse™ (NGC)*, sponsored by the Agency for Healthcare Research and Quality (AHRQ), U.S. Department of Health and Human Services.

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