Disease/Medical Condition

SICKLE CELL DISEASE
(also known as “SCD”, “hemoglobin S disease”, and “Hb S disease”; term encompasses sickle cell anemia [SCA, which is also known as hemoglobin SS disease], sickle cell hemoglobin disease [hemoglobin SC disease], sickle beta-thalassemia [hemoglobin Sβ that], hemoglobin SD disease, and hemoglobin SE disease)

Date of Publication: April 12, 2016

Note: This fact sheet focuses on sickle cell anemia unless otherwise indicated. Sickle beta-thalassemia is further addressed in the thalassemia fact sheet.

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

- Is medical consult advised? Yes, liaison with the patient/client’s hematologist (blood specialist) or experienced internist, pediatrician, or family physician is advisable regarding the severity and management of the patient/client’s sickle cell disorder before undertaking dental hygiene treatment for the first time. Medical consult is also warranted for suspicious, but as yet undiagnosed, anemia or hemoglobinopathy (e.g., suggestive orofacial features); and if known disease is poorly controlled. Patients/clients of black African ancestry should be screened for sickle cell disease/trait.

- Is medical clearance required? No, for SCD diagnosis. Medical clearance may also be required if patient/client is being treated with medications associated with immunosuppression +/- increased risk of infection (e.g., corticosteroids such as prednisone).

- Is antibiotic prophylaxis required? Yes, if there is compromised immunity, which places patient/client at risk for distant-site infection from transient bacteremia resulting from invasive procedures. As well, antibiotic prophylaxis is often recommended for major dental surgical procedures to reduce the risks of wound infection and osteomyelitis (even though the evidence supporting such use is largely lacking). Furthermore, daily penicillin is often prescribed until age 5 or 6 years to prevent serious infection in children with SCD; some adults also take ongoing antibiotic prophylaxis, particularly if they have had their spleen removed.

- Is postponing treatment advised? Yes, until the patient/client has been medically cleared, including reducing propensity for a sickling crisis (e.g., infection treated and adequate hydration ensured), addressing of severe anemia, and ascertaining if antibiotic prophylaxis is warranted. To minimize complications, hemoglobin levels should be above 110 g/L, and the patient/client should be free from symptoms of anemia. Patients/clients who are short of breath and in whom Hb levels are less than 110 g/L, abnormal heart rate is present (e.g., tachycardia), or oxygenation is less than 91% by pulse oximetry are medically unstable; routine dental hygiene treatment should be deferred until health status improves. If a sickle cell crisis is suspected, emergency protocol should be initiated and prompt transfer to an emergency department is indicated. Patients/clients with heterozygous sickle cell trait (see below), as distinct from homozygous sickle cell disease, are not at risk for adverse events during dental hygiene treatment unless severe hypoxia, severe infection, or dehydration also is present.

Is the initiation of invasive dental hygiene procedures contra-indicated? Yes. This is a blood disorder that may affect appropriateness or safety, and scaling and root planing, including curetting surrounding tissue, are contraindicated until the patient/client is medically cleared. Other potential contraindications include: spleen damage or splenectomy (resulting in compromised immunity and thus increased risk for serious infections); certain medication side-effects (e.g., reduced white cell count [compromised immunity] or reduced platelet count [abnormal bleeding risk] resulting from hydroxyurea treatment); and bone marrow transplantation (because it variously involves chemotherapy or radiotherapy to destroy the host’s bone marrow followed by a life-long regimen of immunosuppressive therapy to prevent rejection).

- Is medical consult advised? See above. Patient/clients with sickle cell anemia may be in danger of a sickle crisis if the disease is not detected and managed before invasive procedures are started.

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Oral management implications

- All black Canadian patients/clients (i.e., those of black African ancestry) should be asked about the presence of sickle cell disease in their family.
- Good oral hygiene and preventive care are very important, because oral infection can lead to a sickle cell crisis and/or osteomyelitis.
- Patients/clients with sickle cell disease can safely receive routine dental hygiene care during non-crisis periods. However, long procedures should be avoided; appointments should be kept short to reduce stress.
- A significantly elevated temperature (38°C or more) should prompt postponement/cessation of dental hygiene procedures and immediate referral for medical follow-up (e.g., hospital emergency department).
- Adequate fluid intake should be ensured pre-, intra-, and post-appointment to avoid dehydration.
- Intravenous sedation, if applicable, must be used very cautiously. Strong narcotics and barbiturates should be avoided because respiratory suppression leads to hypoxia and acidosis, which may precipitate an acute sickle crisis.
- Pulse oximetry monitoring is prudent during invasive dental hygiene procedures; oxygen saturation should be above 95%.
- High doses of salicylates (e.g., acetylsalicylic acid, or ASA) should be avoided for oral pain management, because of the potential for acidosis that can trigger a sickle crisis. Acetaminophen, with or without small doses of codeine, may be used for pain control. Non-steroidal anti-inflammatory drugs (NSAIDs) should be used cautiously (and for a few days only), if at all, due to risk of exacerbating underlying nephropathy (kidney disease).
- If infection occurs post-procedure, a dentist and/or physician must treat it expeditiously. Cellulitis may require hospitalization for monitoring and intravenous antibiotic administration.
- Management of a sickle cell crisis is largely supportive and directed at symptoms; it includes the administration of oxygen and intravenous and oral fluids. A sickle cell crisis is a medical emergency and necessitates initiation of emergency protocol.

Oral manifestations

- Delayed eruption of teeth and dental hypoplasia (including disorders of enamel and dentine mineralization) are common in patients/clients with SCD. There may also be changes to the superficial cells of the tongue (“smooth tongue”), malocclusion, hypercementosis, and severe periodontitis (particularly in children).
- Bone marrow hyperplasia in the maxillofacial bones can result in characteristic, though relatively uncommon, facial findings in children with SCD. These include mid-facial overgrowth, frontal bossing, and protrusion of the maxilla (which exposes the teeth, depresses the nasal bridge, and causes malocclusion).
- Mucosal pallor and/or jaundice1 (particularly yellow discolouration of the gingiva) may arise from the hemolytic anemia associated with sickle cell disease.
- Toothaches are more frequent in patients/clients with sickle cell anemia, often secondary to persistent hypoxia of the dental pulp and resultant pulpal necrosis. Pulp pain may be mimicked by infarction/thrombosis of dental pulp vessels, which exposes vital pulp to hypoxia. In some patients/clients, pulpal necrosis may be asymptomatic, which contributes to extensive tissue damage before detection.
- Dental caries is a common finding in patients/clients with SCD, particularly in persons of lower socioeconomic status.

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1 Jaundice is caused by hyperbilirubinemia resulting from large-scale destruction of red blood cells.
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Oral manifestations (cont’d)

- Periodontal disease tends to be associated with lack of oral hygiene maintenance, rather than SCD itself. However, gingival enlargement can occur secondary to vaso-occlusive events and hemorrhage, with fibrous repair, resulting from erythrocyte sickling.
- Mental nerve neuropathy (resulting in numbness in the lower lip and chin, hence the term “numb chin syndrome”) can result from vaso-occlusive events. In some persons, teeth may also exhibit loss of sensation (when other branches of the inferior alveolar nerve are involved), and patients/clients may bite their lip unintentionally.
- Osteomyelitis of the jaw secondary to SCD is rare; however, when it occurs, the mandible is most commonly affected facial bone. Mandibular osteomyelitis typically occurs in the third decade of life, whereas maxillary osteomyelitis tends to strike during the first decade. Persistent ischemia (caused by cell sickling) induces bone necrosis, which predisposes to secondary infection.
- Dental radiographs may show enlarged, irregular bone medullary spaces associated with bone marrow hyperplasia (due to erythrocyte production response to RBC destruction); increased widening and decreased numbers of trabeculations (particularly in alveolar bone); loss of height of alveolar bone; and osteoporosis (particularly thinning of the inferior border of the mandible). Bone appears more radiolucent, with prominent striations; the trabeculae between teeth may take on a coarse horizontal row or “staircase” pattern. Radiopaque areas caused by repairs to bone infarction may be present in the maxilla and/or mandible.
- Mucositis may be present in patients/clients undergoing chemotherapy or radiation therapy related to bone marrow transplantation.
- Xerostomia is a common side-effect of some medications (e.g., duloxetine, gabapentin, and amitriptyline) used to manage chronic pain associated with SCD.

Related signs and symptoms

- Sickle cell disease (SCD) is a group of inherited red blood cell (RBC) disorders. Persons with SCD have abnormal hemoglobin, called hemoglobin S or sickle hemoglobin2, in their RBCs. When a person has two hemoglobin S genes (HbSS), the disease is referred to as sickle cell anemia (SCA), which is the most common and severe form of sickle cell disease. Hemoglobin SC disease and sickle beta-thalassemia (hemoglobin Sβthal) are two other forms of SCD.
- SCD is inherited in an autosomal recessive manner. This lifelong condition is found predominately in persons of African (black), Asian (Indian), Middle Eastern, Mediterranean, and Hispanic descent, and it is more common in regions of malaria endemicity.
- In Canada, an estimated 5,000 persons have SCD, and up to 1 in every 2,500 babies will be born with it. Nearly 150,000 persons are carriers of the trait.
- SCD causes problems in two ways: (1) by the breakdown of red blood cells (hemolysis) and (2) by blocking the flow of blood in the blood vessels (vaso-occlusion). This often causes pain, especially in the bones.
- The erythrocytes (red blood cells, or RBCs) in SCD become sickle-shaped when blood experiences lowered oxygen tension or decreased pH, or when the patient/client becomes dehydrated. This leads to increased blood viscosity, increased adhesion of RBCs, reduced blood flow, hypoxia, vascular occlusion, and further sickling. Lifespan of RBCs is reduced. Exertion, exercise, administration of a general anesthetic, pregnancy, and even sleep can trigger sickling of RBCs in persons with sickle cell disease. However, sickling crises are rare in persons with sickle cell trait.

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2 Normal red blood cells contain hemoglobin A.
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Related signs and symptoms (cont’)

- Persons with sickle cell trait (SCT) are generally asymptomatic unless they experience situations in which there are abnormally low concentrations of oxygen (e.g., during administration of general anesthesia, in an unpressurized airplane, etc.).
- Diagnosis of SCD includes the use of red blood cell indices and a deoxygenation test, along with confirmatory electrophoresis or chromatography. Sickle cells may be seen on a blood smear. Hemoglobin (Hb) level³ and erythrocyte count are typically reduced.
- All newborns in Ontario are specifically screened for sickle cell anemia, hemoglobin SC disease, and sickle beta-thalassemia on their newborn blood screening test. Some other hemoglobinopathies may also be detected as part of this screen.
- Sickle cell anemia nearly always presents before age 30 years. All tissues and organs of the body can be affected, and systemic complications occur especially in areas that are most compromised by hypoxia and infarction. The disease presents with variable clinical manifestations, and different degrees of severity dependent on the stage at which the disease is first detected, the patient’s age, number of hospitalizations, the need for blood transfusions, and the need for continuous drug use (e.g., hydroxyurea and/or pain medications).
- Signs/symptoms of SCD are the result of anemia and small blood vessel occlusion. Common manifestations include weakness, pallor, jaundice, shortness of breath, fatigue, and nausea. Other sequelae are dactylitis (painful swelling of hands and feet), organomegaly (enlargement of internal organs, particularly the heart, liver, and spleen), cardiac failure, abdominal pain, and delays in growth and puberty. In the hemolysis-associated form of SCD, icterus (yellow colour of the sclera of the eyes), leg ulcers, and pulmonary hypertension are prominent features. The vaso-occlusive form is characterized by chest pain (acute chest syndrome), stroke, avascular necrosis of joints (particularly the hips and shoulders), and priapism (painful, prolonged penile erection).
- SCD can cause nephropathy (kidney damage), leading to frequent urination, bedwetting, and/or nocturnal enuresis (uncontrolled urination during the night). Hematuria (blood in urine) may also occur.
- Many adolescents and adults with SCD suffer from chronic pain, which differs from crisis pain and organ damage pain, and whose cause is not well understood. Associated anxiety or depression is common.
- Three severe systemic manifestations of sickle cell anemia are sickle cell crisis (in which there is severe sickling of erythrocytes resulting in vaso-occlusion), acute chest syndrome (in which there is lung involvement), and aplastic crisis (in which production of RBCs stops).
- A sickle cell crisis is a reversible, extreme pain episode caused by deprivation of blood and oxygen to tissues. It can be precipitated by stress, illness/infection, temperature changes, dehydration, being at high altitudes, and pregnancy.
- Acute chest syndrome manifests as chest pain, wheezing, cough, fever, and hypoxia.
- An aplastic crisis can occur when a person (usually a child) with sickle cell disease is infected with parvovirus B19, the virus that causes fifth disease in children. This acute illness, in which severe anemia results, may also be less frequently triggered by Epstein Barr virus or streptococcal infection hypoxia, hypersensitivity reactions, acidosis, trauma, or dehydration.
- Most persons with SCD usually have mild to moderate anemia. At times, however, they can have severe anemia, which may be life threatening. In addition to infection-triggered aplastic crisis, severe anemia in an infant or child may be caused by a splenic sequestration crisis, in which red blood cells get stuck in the spleen.

³ Anemia is generally defined as Hb level less than 120 g/L in women and less than 130 g/L in men.
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Related signs and symptoms (cont’)

- Acute chest syndrome and ischemic stroke tend to strike patients/clients with SCD aged from birth to 20 years, whereas osteonecrosis of the hip and shoulder joints, priapism, liver disease, and gallstones tend to manifest in persons aged 20 to 40 years. In persons older than 40 years, pulmonary hypertension, nephropathy, retinopathy (damage to retina of the eye), heart murmurs, and sudden cardiac death due to arrhythmias are possible complications. Hemorrhagic strokes are more common in adults than children with SCD.
- Sickle cell leg ulcers (which may heal and then recur) tend to appear in SCD patients/clients aged more than 10 years.
- Repeated infarctions in the spleen can lead to hypofunction, with resultant compromised immunity and susceptibility to infections.
- Pregnancies in women with SCD are risky for both the mother and the fetus/baby. Mothers may have medical complications including infections, blood clots, and high blood pressure. There is also elevated risk of miscarriage, premature birth, and underweight babies.
- Many complications of SCD can be prevented or reduced by either regular blood transfusions or daily oral hydroxyurea. However, chronic blood transfusions can result in iron overload; complications include impairment of heart, lung, and liver function. Hydroxyurea (which increases fetal hemoglobin) can cause leukopenia (low white blood cell count) or thrombocytopenia (low platelet count), and, in rare cases, it can worsen anemia.
- The only current cure for SCD is hematopoietic stem cell transplantation (i.e., bone marrow transplant), which is not a viable option for most people.
- Up to 25% percent of people with hemoglobin SS (sickle cell anemia) and 10% of people with hemoglobin SC suffer a clinical stroke by age 45 years.
- Untreated sickle cell anemia may result in early childhood death. In North America, the life expectancy of a person with SCD has increased in recent decades to about 40 to 60 years.

References and sources of more detailed information

- Sickle Cell Disease Association of Canada [http://www.sicklecelldisease.ca](http://www.sicklecelldisease.ca)
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References and sources of more detailed information (cont’)

- National Heart, Lung, and Blood Institute of the National Institutes of Health (U.S.)
  http://www.nhlbi.nih.gov/health/health-topics/topics/sca
  http://www.nhlbi.nih.gov/health/health-topics/topics/sca/treatment#HU
- Newborn Screening Ontario http://www.newbornscreening.on.ca/bins/content_page.asp?cid=7-21-350
- Indiana Hemophilia and Thrombosis Center
  http://www.aapd.org/media/policies_guidelines/g_antibioticprophylaxis.pdf

* 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: September 27, 2015