

Black History Month Webinar Series

Sickle Cell Anemia in Primary Care- A primer on screening, diagnosis and counselling

Dr. Eniola Salami

Sickle Cell Disease (SCD) in Alberta

- 1 in 128 infants born in Alberta will be born with sickle cell trait
- Predominates in African, Mediterranean, Middle Eastern, Asian communities
- Prevalence in some communities can be as high as 1 in 400
- 6500 people across Canada have this condition

When to test

- Alberta has a newborn screening program for Sickle Cell Disease, however not all infants with SCD will be detected through the screening program.
- Due to the HbF → HbS transition, most infants don't manifest symptoms until 6 months of age.
- Symptomatic screening is essential for infants in target populations born in Alberta
- Currently, all positive screens are referred to the on call pediatric hematologist who then contacts the primary care provider for the infant.

Sickle Cell Disease

- Sickle Cell Disease (SCD) is an autosomal recessive condition - carrier-carrier couples have a 1 in 4 risk of having a child with SCD
- SCD is caused by the mutation of the beta-globin gene resulting in hemoglobin S formation and the creation of "sickled" red blood cells.
- Sickle Cell Trait (SCT) refers to individuals who have one affected beta-globin gene mutation, these individuals are often asymptomatic.
- As per the 2008 SOGC Guideline:
 - Patients from endemic regions/ethnic groups should be screened pre-conception with hemoglobinopathy screen & Complete blood count
 - If both partners screen positive for sickle cell trait they should be referred to genetic counselling**
 - Prenatal diagnostic testing should be offered if patient presents pregnant prior to screening

Blood Testing

- Complete Blood Count with Differential
- Hemoglobinopathy Investigation Panel - tests HbA, HbA2, HbF, HbS, and HbC percentages
- Lactate, haptoglobin, peripheral blood smear
- Reticulocyte Count
- Bilirubin - conjugated and unconjugated
- If febrile - Blood Cx, Urine Cx
- AST, ALT, INR, Albumin
- Serum Creatinine, Sodium, Potassium
- Imaging - +/-CXR Pa & Lat, Transcranial Doppler/MRI → starts at 2. years (CVA)

COMMON FIRST PRESENTATIONS

Anemia

Chronic Anemia, hemolysis, reticulocytosis, aplastic crisis

Acute Chest Syndrome

Pneumonia like illness, fever, chest pain, severe cough, hypoxia

Jaundice

Yellowed Conjunctiva, yellowed skin

Splenic Pooling

Left sided abdominal pain, palpable spleen, hemodynamic compromise/hypotension (EMERGENT)

Pain Crisis

Pain and swelling to limbs, fingers, toes, can have vascular compromise as well**

(Stanford Children's, 2025; Ruchen, 2012; Onimoe & Rotz, 2020)

Where to refer?

- Early referral is critical.
- In Northern Alberta refer to the Pediatric Comprehensive Sickle Cell Program that offers wrap around care, clear protocols for ER presentations, nurse practitioner support and parental support.

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Primary Care Management is centered on:

- 1) Infection Prevention
- 2) Pain Management
- 3) Maintenance of disease modifying therapies

THERAPY

Disease Modification

- Hydroxyurea* - often combined with erythropoietin, functions to increase HbF. Can cause chronic leg ulcers, headaches, changes to integument, GI distress
- L-Glutamine - for > 5 years of age; reduces morbidity and hospitalization; can cause nausea, joint pain, fatigue
- Crizanlizumab-tmca - for > 16 years, monthly infusion, prevents cell adhesion
- Blood Transfusions*
- Bone Marrow Transplant* (potentially curative)

Complication Prevention

- Immunizations* - CRITICAL for Sickle Cell patients who have decreased splenic function
 - H. Influenzae
 - Meningococcal
 - Pneumococcal - broad spectrum
 - Hep B
 - Tetanus
- Antibiotic Prophylaxis - daily prophylactic Penicillin Vk or Amoxicillin from 2 months - 5 years of age, lifelong if spleen removed

U.S. Department of Health and Human Services, 2024; CanHaem 2018

Monitoring

- SCD patients require frequent monitoring with an eye to end organ complications
- Having a systematic approach can help ensure important assessments are not missed

Take Home Message

- Sickle Cell Disease is common in the Alberta Context
- Early Assessment and Detection is essential for reduction of morbidity and mortality
- Common presentations can guide clinical assessments and help shape clinical suspicion
- Continued monitoring for end organ complications and support for chronic pain management are critical roles for the primary care physician
- Persons who can become pregnant and their partners need prompt and effective screening

Resources for patients

- [Ontario Health Sickle Cell Disease guide for patients](#)
- [ICANCOPE App for patients to manage pain crises](#)



Consider using [MyL3Plan](#), a free online tool developed by the Office of Lifelong Learning (L3) that can be used to meet and support the 3 activities/action plans required by the PPIP-CPSA and earn up to 36 Mainpro+ certified credits.

Chronic Pain Management

- Sickle Cell Anemia is a painful condition and patients frequently seek emergency care for pain crises
- Patients with SCD face significant stigma when seeking pain relief during crises
- SCD sufferers are often mislabeled as “drug seeking” and this can lead to delays in proper care
- Opioid analgesia is the first line therapy for sickle cell related pain
- Limited evidence for the use of Tricyclic antidepressants, Gabapentin, and SNRI for management of SCD related pain

Sickle cell disease: Recommended screening and interventions

Nephropathy

Screen annually for albuminuria: spot urine test to estimate protein-to-creatinine ratio
If micro- or macroalbuminuria is present: 24-hour urine test
If protein excretion rate > 300 mg/24 hours, refer to a nephrologist
Consider angiotensin-converting enzyme inhibitor therapy

Pulmonary

Assess for respiratory problems
Pulmonary function testing
If findings suggest pulmonary hypertension, refer for cardiology evaluation

Hypertension

Screen; treat to $\leq 130/80$ mm Hg⁺

Retinopathy

Refer to an ophthalmologist for a dilated eye examination⁺; rescreen in 1–2 years if normal
Refer to a retinal specialist for suspected retinopathy

Stroke

Screening limited to children
Blood transfusion: simple or exchange
Hydroxyurea⁺

Leg ulcers

Inspect lower extremities for active and healed ulcers
Treat with debridement, wet-to-dry dressings, topical agents
Chronic recalcitrant deep leg ulcers: evaluate for osteomyelitis, consult wound care specialist

Reproductive counseling

Reproductive life plan
Refer partners for hemoglobinopathy status testing if status is unknown
Test women anticipating pregnancy for red blood cell alloantibodies
Discuss contraception choices with no restrictions for use in sickle cell disease: progestin-only contraceptives, barrier methods; reinforce the need for barrier methods for patients on hydroxyurea

Avascular necrosis

Elicit from history and physical examination
Confirm with radiography and magnetic resonance imaging
Refer for physical therapy, orthopedic clinic

- [Skip Action Plan for ER management](#)
- [CanHaem Resources for Patients and Healthcare providers](#)

[Learn more here!](#)