APPHON 2025 Sickle Cell Education

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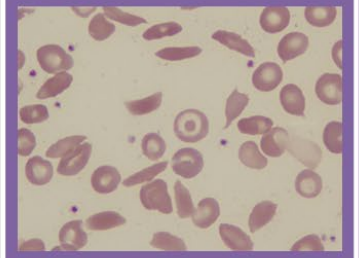
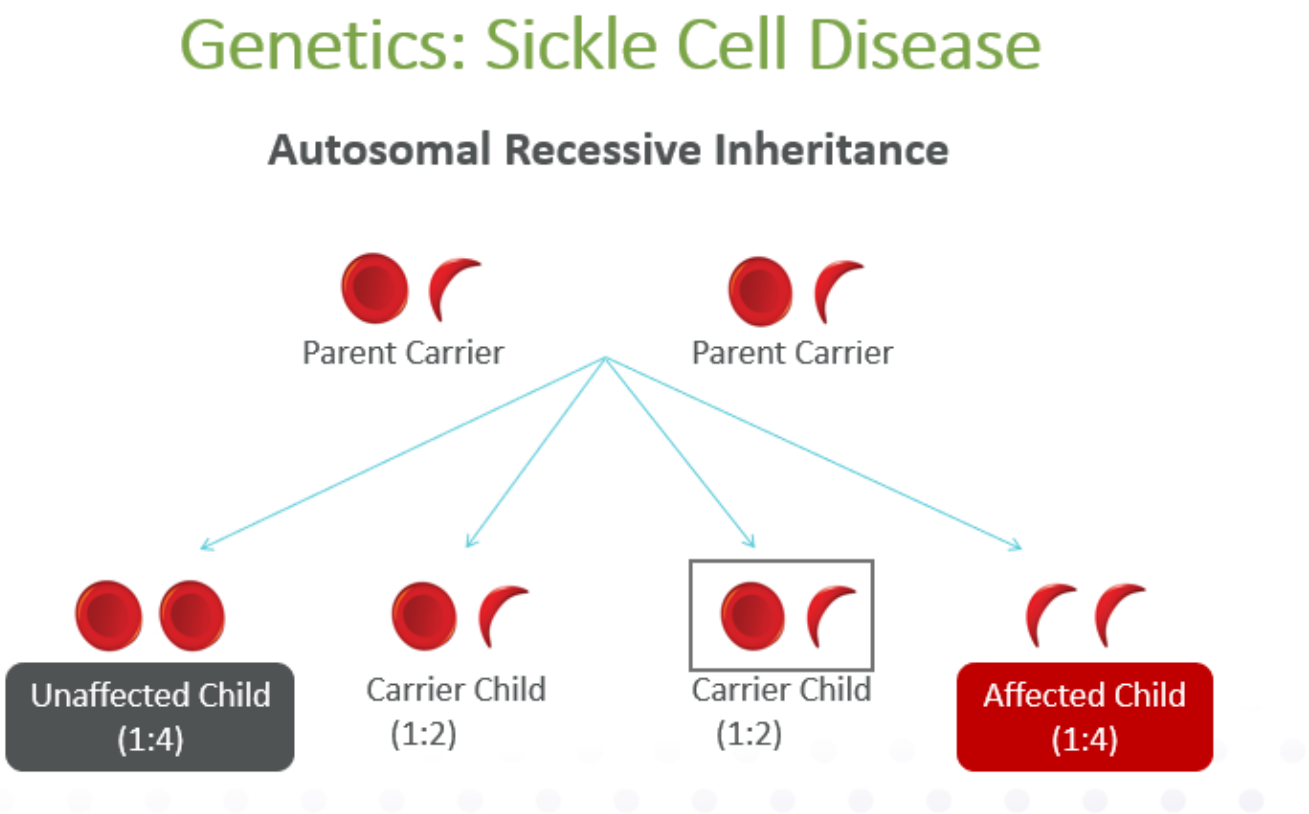
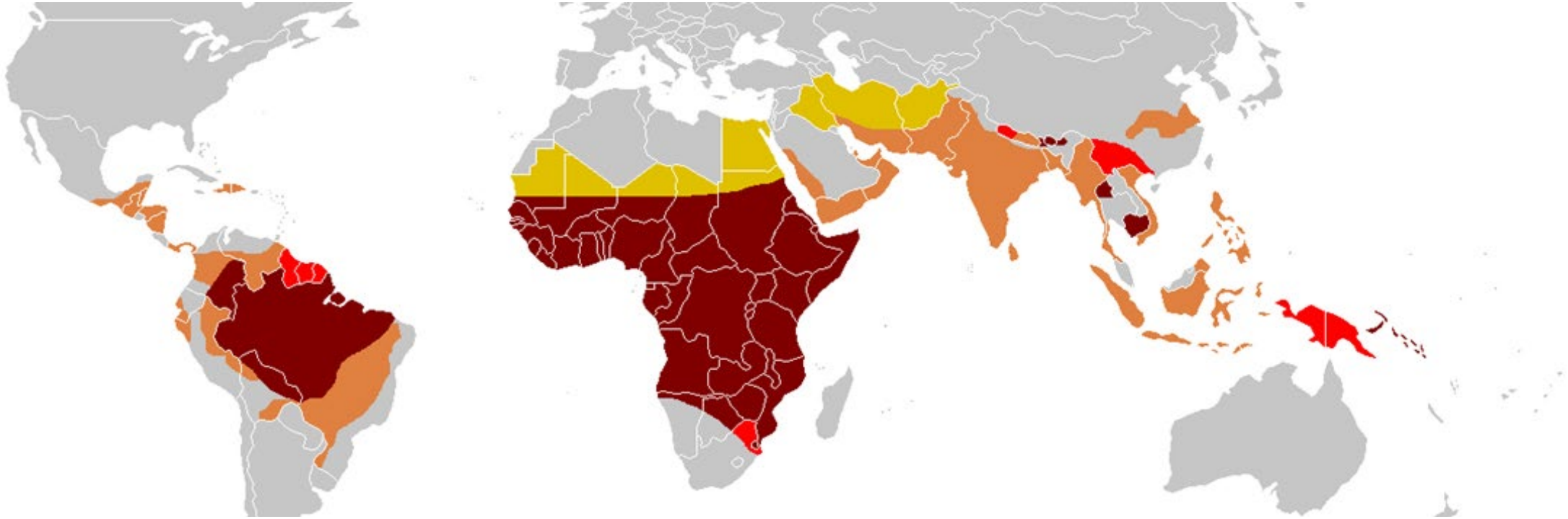
March 19,2025

Sickle Cell Disease

Definition	Chronic disease with acute exacerbations Caused by mutation in DNA that determines HGB resulting in absence of normal HbA
RBC effect 	Normal RBCs are flexible, biconcave discs with lifespan of 120 days SCD RBCs polymerize, forming microtubules making cells crescent-shaped and friable with 10–20-day lifespan Leads to vaso-occlusion
Genetics	Autosomal recessive
Presentation	Newborn screening Apparent by six months Pallor/Jaundice Acute/Chronic complications

Genetics of Sickle Cell Disease





Epidemiology

- Described in 1904 by James Herrick
- Most common inherited disorder in the United States
- Prevalence follows the Malarial belt, higher prevalence in African, Caribbean, Mediterranean, South America, Southeast Asia

Canadian Statistics

- An estimated 6,000 Canadians are living with sickle cell disease.
- About 1 in 2500 newborns in Canada will have sickle cell disease.
- All provinces have a newborn screening program.
- NL, MB & SK do not screen for hemoglobinopathies.
- In the Maritimes we have approx. 73 pediatric patients with sickle cell disease.
- 42 patients in New Brunswick
- 28 patients in Nova Scotia
- 3 patients in PEI
- 17 patients in NL

Types of Sickle Cell Disease

Genotype	Predicted Severity
HbSS	Moderate to very severe
HbSC	Mild to severe
HbS-β^0 (zero) thal	Moderate to very severe
HbS-β^+ (plus) thal	Mild to moderate
HbS-D (Punjab) HbS-O Arab	Moderate to very severe

ED Sickle Cell Pathways

All preprinted orders and clinical care pathways for the management of sickle cell disease can be found on the APPHON website at:
https://www.apphon-rohppa.com/en/guidelines/sickle-cell-asplenia-guidelines

Initial Management of Sickle Cell Disease (SCD) – ED Pathway

Triage and Assessment

A child with SCD presenting to ED with fever or pain should be assessed within 30 minutes of arrival.

Check for signs of complications,

- Vaso-occlusive crisis (painful crisis)
- Fever – sepsis
- Acute chest syndrome
- Stroke
- Priapism
- Aplastic crisis
- Acute splenic sequestration.

General Sickle Cell Management

1. **Pain**: Start analgesics promptly – treat pain aggressively.
   - Mild: acetaminophen, ibuprofen.
   - Moderate: Consider oral morphine or hydromorphone.
   - Moderate to severe: morphine or hydromorphone IV, repeat as needed (refer to IWK DDG for intermittent IV dosing), may need continuous infusion. Refer to Sickle Cell Disease Pain Admission Orders on APPHON website.
2. **Fluids**:
   - Encourage oral fluids. May require IV fluid bolus 10 – 20 mL/kg.
   - Avoid excess fluids to reduce risk of chest crisis.
   - Do **not** delay starting IV fluids or analgesics for topical anaesthetic cream. If unable to obtain IV access after 2 attempts, consider ultrasound guided IV placement or page anesthesia/anesthesia assistant.
3. **Antibiotics:** If patient presents with fever start IV Ceftriaxone. Refer to guidelines on APPHON website.
4. **Consultations:** May require blood transfusion – discuss with Hematology. Early PICU review and respiratory support if concern for acute chest crisis.

Contact Hematologist on call

Investigations:

- CBCD including reticulocyte count.
- Blood group & cross match.
- CRP, blood and urine cultures if febrile.
- Serum Creatinine, BUN and electrolytes and LFTs if dehydrated or jaundice.
- Chest X-ray if respiratory symptoms.
- Other imaging as required.

Vaso-occlusive crisis (painful crisis)	Fever – sepsis	Acute chest syndrome	Stroke	Acute splenic sequestration	Aplastic crisis	Priapism
Precipitated by dehydration, hypoxia or infection. All episodes of pain should be treated initially as vaso-occlusive disease as per Sickle VOC clinical pathway on APPHON website. Chest pain may indicate an acute chest syndrome rather than as a vaso-occlusive episode if associated with respiratory symptoms. *Refer to clinical care pathway on the APPHON website.*	Patients are functionally asplenic and at greater risk for invasive disease by encapsulated organisms. **Specific management:** • Start IV Ceftriaxone as per the ED SCD fever orders on APPHON website. • Consider coverage for atypical organisms (Clarithromycin) if significant respiratory component. • Obtain appropriate cultures ○ Blood, sputum, urine. If pain is also present, treat as vaso-occlusive crisis. If cough or dyspnoea is present, look and treat for acute chest syndrome.	Life threatening condition. Suspect if respiratory distress, hypoxia or chest pain. **Specific management:** • Oxygen to keep oxygen saturations > 94% or for comfort. • Analgesia as above. • Start IV antibiotics – Ceftriaxone and Clarithromycin as per the ED SCD fever orders on APPHON website. • Chest X-ray – but don't delay treatment. • Consider simple transfusion in consultation with hematology. Do NOT exceed a post transfusion Hb of 100g/L. • Early referral to PICU for respiratory support if significant hypoxia or respiratory distress. *Refer to clinical care pathway on the APPHON website.*	Can occur suddenly or as a complication of acute chest syndrome or aplastic crisis. **Specific management:** • Neuroimaging required to determine if hemorrhagic or ischemic stroke. • MRI is modality of choice. (ED or hematology to order). If not available, • CT - NO CONTRAST (risk of hyperviscosity). **Transfusion support:** • Options include initial simple transfusion to Hb 100 g/L followed by red cell exchange. *Refer to clinical care pathway on the APPHON website.*	Anemia (↓Hb >20g/L) with thrombocytopenia and acute splenomegaly. May present acutely shocked. **Specific management**: • Fluid resuscitation – NS 0.9% 10-20 mL/kg initial transfusion to aim for Hb of 50-60 g/L initially to ameliorate hemodynamic instability. • Do not increase Hb by > 30 g/L of presenting Hb with initial transfusion and do NOT exceed a post transfusion Hb of 100 g/L. Auto-transfusion may occur if hemoglobin is increased excessively or too quickly. This increases risk of stroke due to hyperviscosity. • IV antibiotics if febrile as per ED SCD fever orders on APPHON website. *Refer to clinical care pathway on the APPHON website.*	An acute illness with decreased hemoglobin without a reticulocyte response (usually <1%). Usually associated with acute infection including parvovirus. Present with pallor +/- shock. **Specific management:** • Intravenous fluids and oral intake to a total of maintenance. • Transfuse red blood cells if patient is asymptomatic with anemia or Hb <50 g/L (do NOT increase Hb by > 30 g/L of presenting Hb with initial transfusion). • Start IV antibiotics if febrile – Ceftriaxone as per the ED SCD fever orders on APPHON website.	Two forms – intermittent or prolonged. **Specific management**: • Do not use ice. • Simple measures e.g. moderate exercise, take a bath or shower. • Empty bladder – may need catheter. • Analgesia, oxygen, hydration with alkalization of the urine should be commenced as soon as possible. Consult Pediatric Urologist and on-call hematologist.

Version: January 31, 2025

Clinical Presentation in Pediatrics

Fever

- Patients develop functional asplenia by the ages of 2-4 years.
- Risk of overwhelming sepsis from polysaccharide encapsulated organisms with streptococcus pneumoniae being the most common. Sepsis is the leading cause of death in children with sickle cell disease.
- Fever is a lifelong emergency.
- Penicillin Prophylaxis and supplemental immunizations are recommended for all patients.

APPHON Sickle Cell Treat Promptly Card

Definitions

Fever

- Temperature taken at home by parent **MUST** be taken into account
- Mouth/Ear
  - 38.3°C & over- 1 reading
  - 38°C & over – 2 readings 1 hour apart
- Armpit (Axilla)
  - 37.8°C & over- 1 reading
  - 37.5°C & over – 2 readings 1 hour apart

Immediate assessment:

- Source of infection: consider meningitis, AOM, osteomyelitis, etc.

Patient Information

Please Fax assessment and treatment documents to **902-470-7208**

Name: _____

DOB: _____(dd/mm/yyyy)

Diagnosis: _____

Co-morbidities: _____

Antibiotic Prophylaxis: _____

Other Medications: _____

Prescriber: _____

Date: _____(dd/mm/yyyy)

Guidelines for Emergency Management of ACUTE ILLNESS OR FEVER in Children with Asplenia or Hyposplenia

Treat Promptly!

Atlantic Provinces Pediatric Hematology Oncology Network
Réseau d'Oncologie Hématologie Pédiatriques des Provinces Atlantiques
(APPHON/ROHPPA)

Version Date: May 2021

Fever and/or **acute illness** in children and youth with asplenia or hyposplenia can be *life threatening* and must be *treated promptly*. Overwhelming bacterial infection is a significant risk in patients with no splenic function or absent spleen (asplenia) or a dysfunctional spleen (functional asplenia/hyposplenia).

Assessment

1. **Triage as a Level 2**
2. Stabilize child
3. Draw CBC, diff, lactate, blood culture stat within **30 mins**
4. Establish vascular access
5. Start antibiotics within **60 mins**
   - **DO NOT WAIT FOR CBC RESULTS**
   - If hemodynamically stable, a **maximum of 3 attempts** to insert an IV cannula; if unsuccessful, IM ceftriaxone should be given using the reconstitution guidelines to include lidocaine (without epinephrine) for those over 5 kg
   - Refer to **pre-printed orders and algorithm** for guidance
6. Referral to nearest emergency department as clinical deterioration can be sudden

START ANTIBIOTICS IMMEDIATELY!

Treatment

Refer to guidelines and use pre-printed orders at www.apphon-rohppa.com

KNOWN ALLERGIES:_____

NOTE: These recommendations do NOT change for those with a penicillin allergy

If meningitis is NOT suspected:

❑ cefTRIAXone 100 mg/kg/dose IV/IM q24h (max 2000 mg/dose)

If greater than 5 years old and suspected atypical pneumonia:

❑ clarithromycin 7.5 mg/kg/dose PO BID (maximum 500 mg/dose)

Suspected meningitis:

❑ cefTRIAXone 100 mg/kg/dose IV x 1 (max 2000 mg/dose), then 12 hours later 50 mg/kg/dose IV q12h (max 2000 mg/dose)

❑ vancomycin

- ***Less than 12 years of age***: vancomycin 15 mg/kg/dose IV q6h (max 1000 mg/dose)
- ***12 years of age and older***: vancomycin 15 mg/kg/dose IV q8h (max 1000 mg/dose)

PPO Fever/ Acute Illness ED Management

Sickle Cell Disease and/or Asplenia with Fever or Acute Illness ED/Clinic Management Greater than 1 month old

K07002307 Jun/7/2002 M
SCA,TEST Visit
ER0000145/12 **HCN:** 22222222
Van den Hof, TEST / TEST, Maureen
Dec/8/2011

Patient:_____

☐Alert Record Reviewed ☐No Allergies Known

☐Allergies–Adverse Reactions–Cautions: _____

Age_____ Patient's Weight_____kg Date of Patient's Weight _____

DIAGNOSIS: _____

Items preceded by a **bullet** (•) are active orders. Items preceded by a **checkbox** ( ) are only actioned if checked (√)
Refer to APPHON website for the link to the CanHaem Consensus Statement on the care of patients with sickle cell disease in Canada (https://www.apphon-rohppa.com/en/guidelines/sickle-cell-guidelines)

LAB/INVESTIGATIONS

Within 30 minutes of arrival:

- Blood culture (ideally before giving antibiotics, but do not delay giving antibiotics beyond 60 minutes from time of arrival)
- CBCD, reticulocyte count, Na+, K+, BUN, creatinine, ALT, AST, bilirubin (total and direct), blood glucose
  - ☐Blood gas, lactate if hemodynamically unstable
  - ☐Urinalysis ☐Urine culture
  - ☐Chest X–ray (AP and Lateral)
  - ☐NPA (PCR) for: ☐Influenza/RSV ☐Extended viral panel (ID approval required)
    ☐COVID19 (if extended viral panel not available)
  - ☐Throat swab for mycoplasma
  - ☐Lumbar puncture
  - ☐Other _____

MONITORING

- If unstable, place on continuous monitor. Otherwise, BP, HR, RR Temp and pulse oximetry every hour until stable, then every four hours
- Keep oxygen saturation above 94%. Apply oxygen and notify most responsible prescriber and respiratory therapist

DIET/FLUIDS

If acute chest syndrome is suspected:
☐NaCl 0.9% (3/4 x maintenance rate; maximum 150 mL/hour)_____mL/hour IV or oral equivalent
Otherwise:
☐NaCl 0.9% (1 1/2 x maintenance rate; maximum 150 mL/hour)_____mL/hour IV or oral equivalent

MEDICATIONS

Start Antibiotics within 60 minutes of arrival at hospital and call Pediatric Hematologist/Oncologist. NEVER delay empiric antibiotic administration. Do NOT wait for CBC results. If patient has had a confirmed anaphylactic reaction to cefTRIAXone, consult patient's pediatric hematologist/oncologist.

For all patients with fever and/or acute illness:
☐cefTRIAXone (100 mg/kg/dose, maximum 2000 mg/dose)_____mg IV/IM* q24h
** If unable to get IV access after 3 attempts or 45 minutes, use IM route for initial dose* (patients greater than 5 kg, the preferred diluent to use for reconstitution for IM injection is 1% lidocaine without epinephrine as per IWK DDG)

If suspected atypical pneumonia and greater than 5 years old (consult ID for children 5 years and under)
☐ADD clarithromycin (7.5 mg/kg/dose, maximum 500 mg/dose)_____mg/PO BID (in addition to cefTRIAXone)

Suspected meningitis:

- ADD vancomycin (in addition to cefTRIAXone)
  - ☐Less than 12 years of age: vancomycin (15 mg/kg/dose, maximum 1000 mg/dose)_____mg IV q6h
  - ☐12 years of age and older: vancomycin (15 mg/kg/dose, maximum 1000 mg/dose)_____mg IV q8h

DISPOSITION

☐Discharge Home with follow–up in 12 to 24 hours after discharge
Appointment date and time_____Location_____
☐Admit/Transfer to_____and refer to APPHON Inpatient Sickle Cell Order Set

DATE (yyyy/MON/dd) Time (24hr/hh:mm) Prescriber Signature Printed Surname/Registration #

DATE (yyyy/MON/dd) Time (24hr/hh:mm) Verified By (Signature) Printed Surname

Note: Page 2 Clinician Information

This is a two-page document.

Algorithm for the Management of Children Greater than 1 month old or with Sickle Cell Disease and/or Asplenia with Fever or Acute Illness

Fever

38° C tympanic/oral OR 37.8° C axillary or greater 1 hour apart or 38.3° C tympanic/ oral or greater

Temperature taken **at home** MUST be considered a true temperature

Immediate Assessment (ABCs)

Source of infection: ?meningitis, AOM, osteomyelitis, etc.

Look for co–morbidities: splenic sequestration, aplastic crisis, etc.

Appropriate cultures + Labs

Administer antibiotics within 1 hour of arrival

Does patient have clinical signs of meningitis?

YES → Perform Lumbar Puncture if clinically appropriate, do not delay antibiotics
Antibiotics: cefTRIAXone (meningitic dosing) + Vancomycin

NO → cefTRIAXone (non–meningitic dosing) → If patient greater than 5 years with suspected atypical pneumonia, add clarithromycin

Discuss with on–call pediatric hematologist–oncologist

Does patient have hypoxia, or cough, shortness of breath, and/or chest pain

Consider Acute Chest Syndrome

- Maintain oxygen saturation over 94%
- Chest X–ray (AP + Lateral)
- Avoid excessive hydration (75% maintenance IV Fluids

Usual organisms include: *Streptococcus pneumoniae, Haemophilus influenzae, Neisseria meningitis, Salmolella spp, and Escherichia coli*

Admission Criteria (list is not exclusive)

Patient Factors	Environmental	Clinical	Investigations
• Age less than 1 year • Prophylaxis indicated but patient not compliant • History of invasive pneumococcal infection • Patient on chronic transfusion therapy for stroke	• No reliable method of contact • Lives more than 45 minutes away from nearest ED • Unable to return in 12 to 24 hours for reassessment	• Temperature greater than 39.5° C • Dehydration • Abnormal vital signs • Toxic appearing • Signs of meningitis • Suspected **acute chest syndrome** • Any concerning features	• WCB greater than 30 x 10^9/L • ANC less than 0.5 x 10^9/L • Hgb less then 60 g/L • Plt less than 100 x 10^9/L • Urinalysis positive for blood, nitrates, or leukocyte estrase • Infiltrates on chest Xray

- If patient has history of penicillin allergy (including anaphylaxis to penicillin), IV cefTRIAXone can still be used safely.
  – Due to the small risk of reaction, observe the patient
- If patient has history of cefTRIAXone allergy or reactions, consult with patient's hematologist/oncologist and/or refer to patient's chart if a pre–made plan is in place

Fever/ Acute Illness Follow Up Orders

Management of Children with Sickle Cell Disease and/or Asplenia with Fever or Acute Illness Greater than 1 month old Outpatient Follow Up

K07002307 Jun/7/2002 M
SCA,TEST Visit
ER0000145/12 **HCN:** 22222222
Van den Hof, TEST / TEST, Maureen
Dec/8/2012

Patient:_____
☐Alert Record Reviewed ☐No Allergies Known
☐Allergies–Adverse Reactions–Cautions: _____
Age_____ Patient's Weight_____kg Date of Patient's Weight _____
DIAGNOSIS: _____
Items preceded by a **bullet** (•) are active orders. Items preceded by a **checkbox** ( ) are only actioned if checked (√)
Refer to APPHON website for the link to the CanHaem Consensus Statement on the care of patients with sickle cell disease in Canada (https://www.apphon-rohppa.com/en/guidelines/sickle-cell-guidelines)

GENERAL
- BP, HR, RR Temp and pulse oximetry
- History and physical examination
- Review all test results ordered in emergency department

LAB/INVESTIGATIONS
- Blood culture and sensitivity if temperature is greater than or equal to 38° C one hour apart or greater than or equal to 38.3° C, or if patient appears unwell

☐CBCD ☐daily frequency_____
☐Reticulocyte Count ☐daily frequency_____
☐Na^+, K^+, BUN, creatinine ☐daily frequency_____
☐ALT, AST, bilirubin (total and direct) ☐daily frequency_____
☐Blood gas, blood glucose, lactate if hemodynamically unwell
☐NPA (PCR) for: ☐Influenza/RSV ☐Extended viral panel (ID approval required)
☐COVID19 (if extended viral panel not available)
☐Throat swab for mycoplasma
☐Other _____

MEDICATIONS

☐ If 18 to 24 hours after initial dose in emergency department/clinic:
cefTRIAXone (100 mg/kg/dose, maximum 2000 mg/dose)_____mg IV/IM x 1 dose

☐ If 12 to 18 hours after initial dose in emergency department/clinic:
cefTRIAXone (50 mg/kg/dose, maximum 2000 mg/dose)_____mg IV/IM x 1 dose

- *If blood culture is negative at 24 hours, and patient is well, cefTRIAXone may be stopped after dose above.*
- For an identified source of infection (acute otitis media, streptococcal pharyngitis, etc...), oral antibiotics may be used at the discretion and decision of the treating clinician. This should be written as a separate prescription.
- In patients greater than 5 kg, the preferred diluent to use for reconstitution for IM injection is 1% lidocaine without epinephrine as per IWK Drug Information Website)

FOLLOW-UP/ASSESSMENT

Location _____ Date:_____Time: _____

DATE (yyyy/MON/dd) Time (24hr/hh:mm) Prescriber Signature Printed Surname/Registration #

DATE (yyyy/MON/dd) Time (24hr/hh:mm) Verified By (Signature) Printed Surname

Supplemental Immunizations

Vaccine Schedules for Children with Sickle Cell Disease and Asplenia

Recommendations for vaccines with pneumococcal, meningococcal, Haemophilus influenza type B, Hepatitis B and influenza vaccines. All other routine immunizations should also be kept up to date. Vaccines should be completed 14 days prior to splenectomy. If this is not possible, vaccines should be given 14 days post splenectomy. If compliance after discharge is not assured, then vaccines should be given before discharge from the hospital even if less than 14 days has elapsed since splenectomy.

TABLE 1. Vaccines recommended according to age at diagnosis of asplenia or hyposplenia

Age at which asplenia determined and Immunizations up-to-date for age	Infant less than 12 months				12 to less than or equal to 24 months	Greater than or equal to 2 years	Supplementary dosing
Pneumococcal vaccine							
PCV13 (Prevnar) (4 doses in young children)	✓ 2 mos	✓ 4 mos	✓ 6 mos	✓ 12 to 15 mos	✓✓ 2 doses 8 weeks apart	✓ 1 dose if no prior PCV vaccine or only prior PCV7 or PCV10	
Pneumococcal polysaccharide vaccine (PPV23)						✓ 1 dose (at least 8 weeks after PVC)	1 additional dose 5 years after first dose of PPV23*
Neisseria meningitidis serogroups A, C, W, Y conjugated vaccine							
Men ACWY-CRM (Menveo) **OR**	✓	✓	✓	✓	✓✓ 2 doses 8 weeks apart	✓✓ 2 doses 8 weeks apart	✓ every 5 years
Men ACWY-T (Nimenrix) **OR**	✓	✓		✓ 12 mos	✓✓ 2 doses 8 weeks apart	✓✓ 2 doses 8 weeks apart	✓ every 5 years
Men ACWY-D (Menactra)			9 mos ✓	✓ 11 mos **AND** ✓ 12 to 15 mos	✓✓ 2 doses 8 weeks apart	✓✓ 2 doses 8 weeks apart	✓ every 5 years
Neisseria meningitidis serogroup B vaccine							
4 component 4CMenB (Bexero) **OR**	✓✓or✓ 2 or 3 doses 8 weeks apart (2-5 months give 3 doses) and (6-11 months give 2 doses) followed by 1 booster dose at least 8 weeks after the last dose AND after 1 year of age.				✓✓ 2 doses 8 weeks apart	✓✓ 2 doses 4 to 8 weeks apart if not previously received	No booster doses recommended
Bivalent MenBFHbp (Trumenba)						✓✓✓ Licensed for persons 10 to 25 years of age; 3 doses (2 doses given 4 weeks apart, with another dose at least 4 months after dose two and at least 6 months after dose one).	No booster doses recommended
Haemophilus influenzae type b vaccine							
	✓	✓	✓	✓ 18 mos	✓✓ 2 doses 8 weeks apart	✓ 1 dose	All patients greater than or equal to 5 years should receive one dose of Hib vaccine regardless of previous vaccinations
Influenza vaccine ➢ 6 mos of age and yearly for all age groups							
Hepatitis B vaccine (only required for patients with asplenia/hyposplenia who are exposed to multiple/chronic transfusions -otherwise follow the provincial program guidance for this vaccine).							
➢ 3 doses	✓ 0 mos	✓ 1 mos	✓ 6 mos		Routine titres are not required. If titres are measured inadvertently beyond the 6 months and are non-immune give one dose andmeasure titre 1 month after the dose – if still non-immune complete the series.		

*No more than 2 lifetime doses of PPV23 should be administered.
Live vaccines are contraindicated in an immunocompromised child.
You can give up to 4 vaccines in the same visit in separate sites.
This table is adapted with permission from the Canadian Pediatric Society Position Statement on Preventing and treating infections in children with asplenia or hyposplenia 2019.
Reference documents used in the development of the above information include the Canadian Immunization Guide & the CanHaem Sickle Cell Disease Consensus Statement 2018.

Pneumococcal vaccine (Pneu C-20)

2-7 months 4 dose schedule (2,4,6 months with a dose at 12-15 months. Min interval between doses is 8 weeks)	#1	#2	#3	#4
7-12 months (2 doses at least 8 weeks apart followed by a 3rd dose at 12-15 months at least 8 weeks after 2nd dose)				
12-24 months (2 doses at least 8 weeks apart)				
Greater than 2 years (1 dose)				
Patients previously immunized with PCV13 or PPV 23 give 1 dose (8 weeks since last PCV 13 and 1 year since PCV23				

Men ACWY

2-11 months (3 doses given 8 weeks apart with another dose given between 12-23 months)
Greater than 12 months (2 doses 8 weeks apart then q 5 years)

#1	#2	#3	#4	Q5 years	Q5 Years

Men B

2-5 months (3 doses at 2,4, and 6 months with a booster administered between 12-23 months and 2 months or more from preceding dose)
6-11 months (2 doses greater than 2 months apart with a booster administered between 12-13 months)
12-23 months (2 doses at least 2 months apart with a booster 12-23 months after primary series)
2-10 years (2 doses at least 2 months apart)
10-17 years (2 doses at least 1 month apart)

#1	#2	#3	#4

HIB

2-6 months (3 doses at 2,4,6 months with a booster at 18 months)
7-11 months (2 doses 2 months apart and 1 booster dose given at least 2 months after pervious dose and administered at or after 12 months of age)
12-14 months (1 dose and 1 booster dose given at least 2 months after pervious dose and administered at or after 12 months of age)
15 months and older (1 dose)
All children over the age of 5 should receive a booster dose even if previously immunized.

#1	#2	#3	#4	#5 (after age 5)

Hep B
0,1,6 months apart

#1	#2	#3

Yearly Influenza and Covid 19 immunizations are also recommended for all Sickle Cell Patients

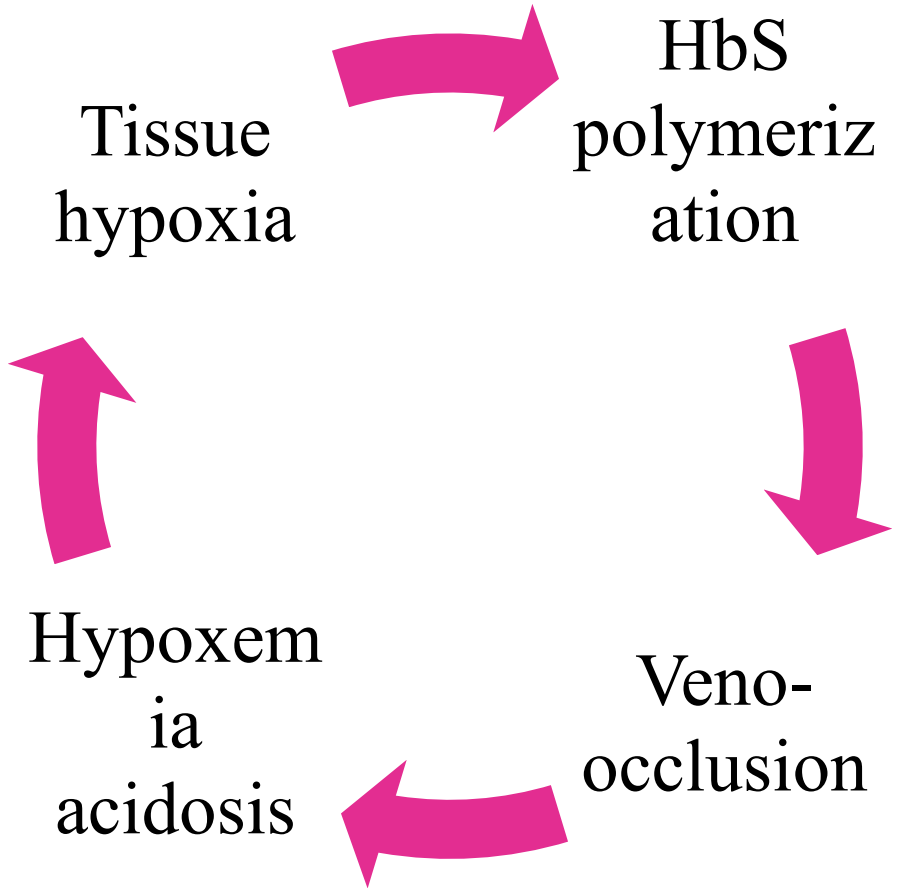
You can give up to 4 vaccines in the same visit in separate sites.

Reference used in the development of the above table include the Canadian Immunization guide (May 2014 and CanHaem Sickle Cell Disease Consensus Statement 2024.

Clinical Presentation in Pediatrics

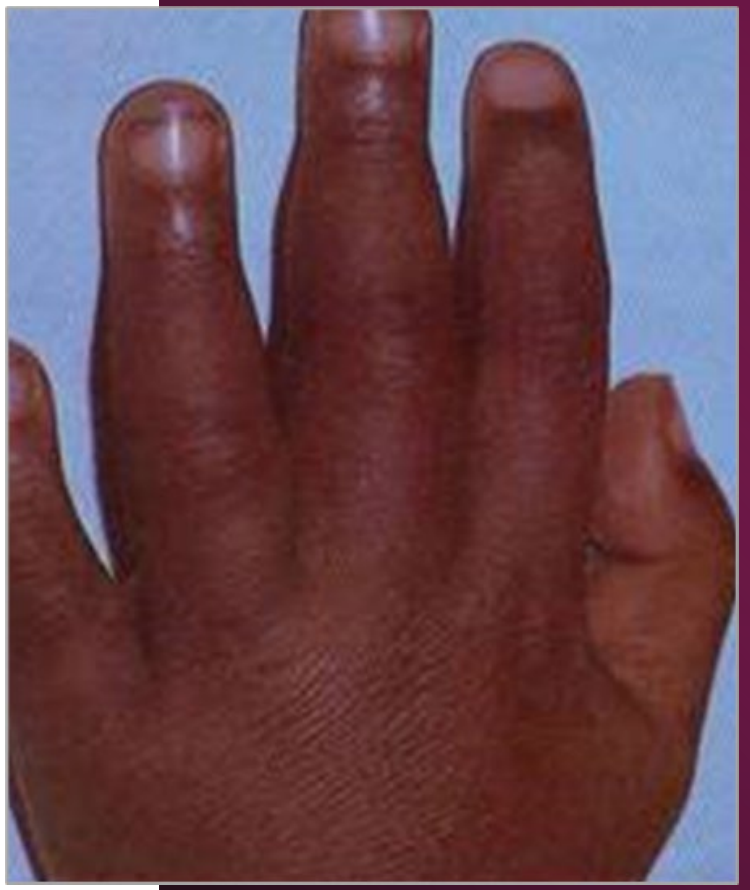
Vaso –occlusive crisis (VOC)

- Fever/ Infection
- Dehydration, Cold or Heat



Vaso- Occlusive Crisis

- Vaso- Occlusive Crisis VOC is the most common reason for a patient with sickle cell disease to seek treatment
- It is the result of three factors
  - Ischemia due to occlusion of the vessels by misshaped RBC’s
  - Endothelia damage
  - Local inflammation



Sickle Cell Disease Pain Admission Orders

Horizon

Sickle Cell Disease Pain Admission Orders
Greater than 6 Months old
HIGH ALERT

*This is a two-page document.

K07002307 Jun/7/2002 M
SCA_TEST Visit
ER0000145/12 **HCN:** 22222222
Van den Hof, TEST / TEST, Maureen
Dec/8/2011

Patient:_____

☐ Alert Record Reviewed ☐ No Allergies Known
☐ Allergies–Adverse Reactions–Cautions: _____
Age_____ Patient's Weight_____kg Date of Patient's Weight _____
DIAGNOSIS: _____
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Refer to APPHON website for the link to the CanHaem Consensus Statement on the care of patients with sickle cell disease in Canada (https://www.apphon-rohppa.com/en/guidelines/sickle-cell-guidelines)

GENERAL
- Admit to_____Admitting Physician:_____

DIET/FLUIDS
☐ Diet as tolerated ☐ Diet _____
- Avoid very cold drinks and caffeine

☐ NaCl 0.9% (maintenance rate; maximum 150 mL/hour)_____mL/hour IV or oral equivalent

MONITORING
- BP, HR, RR, Temp and pulse oximetry every hour until stable, then every 4 hours
- Pain Assessments every 30 to 60 minutes
- Keep oxygen saturation above 94%. Apply oxygen and notify most responsible prescriber and respiratory therapist

☐ Incentive spirometry _____

LAB/INVESTIGATIONS
- Blood culture and sensitivity if temperature is greater than or equal to 38° C one hour apart or greater than or equal to 38.3° C, or if patient appears unwell. If fever, refer to APPHON Sickle Cell Disease and/or Asplenia with Fever or Acute Illness Pediatric Admission Orders

☐CBCD ☐daily frequency _____
☐Reticulocyte Count ☐daily frequency _____
☐Na^+, K^+ BUN, creatinine ☐daily frequency _____
☐ALT, AST, bilirubin (total and direct) ☐ daily frequency _____
☐Blood gas, blood glucose, lactate if hemodynamically unwell
☐Abdominal ultrasound if RUQ pain or epigastric pain
☐Chest X–ray if chest pain or oxygen saturation less than 94% or abnormal breathing
☐Other _____

MEDICATIONS
- Acetaminophen (15 mg/kg/dose, maximum 1000 mg/dose) _____ mg PO q4h PRN(maximum 75 mg/kg/24 hours)
- If greater than 3 months and greater than 5 kg: Ibuprofen (10 mg/kg/dose, maximum 400 mg/dose) _____mg PO q6h PRN for pain(maximum 40 mg/kg/24 hours) **OR**
- If Infant 1-3 months or less than 5 kg: Ibuprofen (5 mg/kg/dose) _____ mg PO q6h PRN

Choose ONE of the following:
☐See completed order set APPHON continuous **Morphine** infusion

If patient has previously received morphine but was not tolerated
☐See completed order set APPHON continuous **HYDROmorphone** infusion

DATE (yyyy/MON/dd) Time (24hour/hh:mm) Prescriber Signature Printed Surname/Registration#

DATE (yyyy/MON/dd) Time (24hour/hh:mm) Verified By (Nurse Signature) Printed Surname

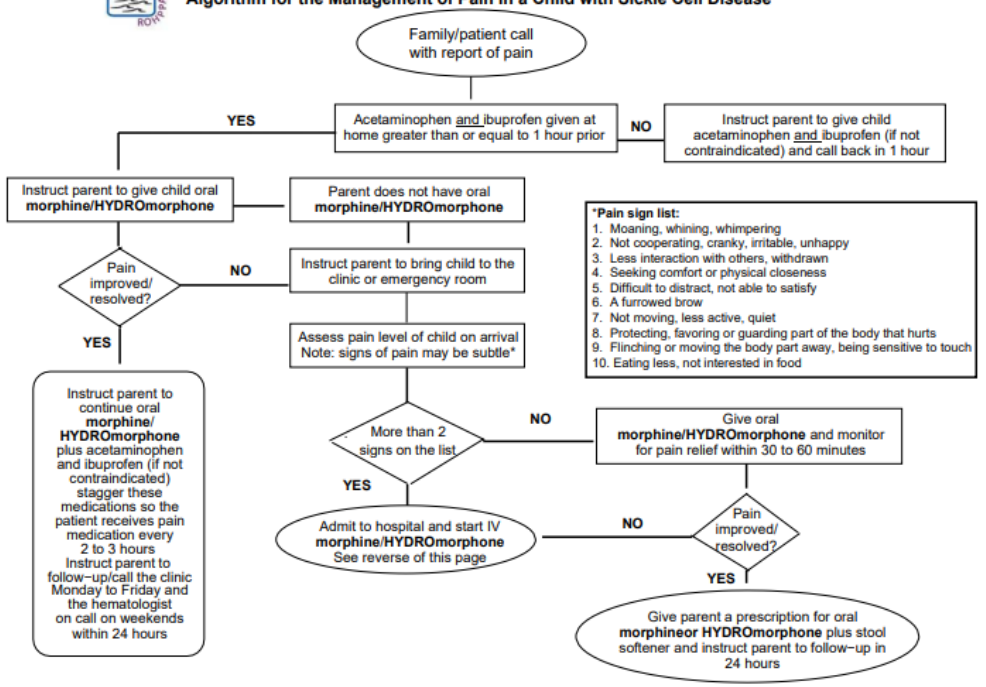
PERMANENT RECORD

Page 2 – clinical information only

APPHON ROHPPA

Algorithm for the Management of Pain in a Child with Sickle Cell Disease

*This is a two-page document.



Pain Management in the ER

Medical emergency

Pain management should commence within 30 minutes

Mild-moderate pain:

- acetaminophen + ibuprofen
- 30-60 min, no relief give morphine

Severe pain:

- Morphine infusion
- Admit to Pediatric unit

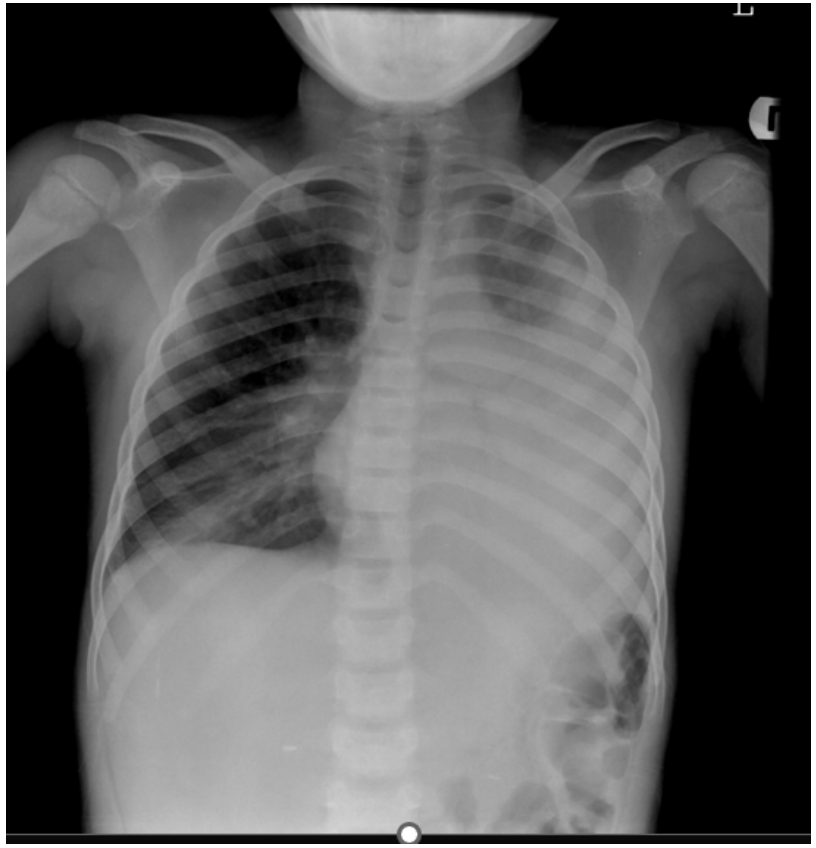
- If abdominal pain assess for splenic sequestration
- If SaO2 < 93%; assess acute chest syndrome; use incentive spirometry
- Maintain hydration: PO/IV
- Offer non-pharmacological pain management strategies
- Discuss with patient/family what has worked in the past
- Keep patient and environment warm

Acute Chest Syndrome

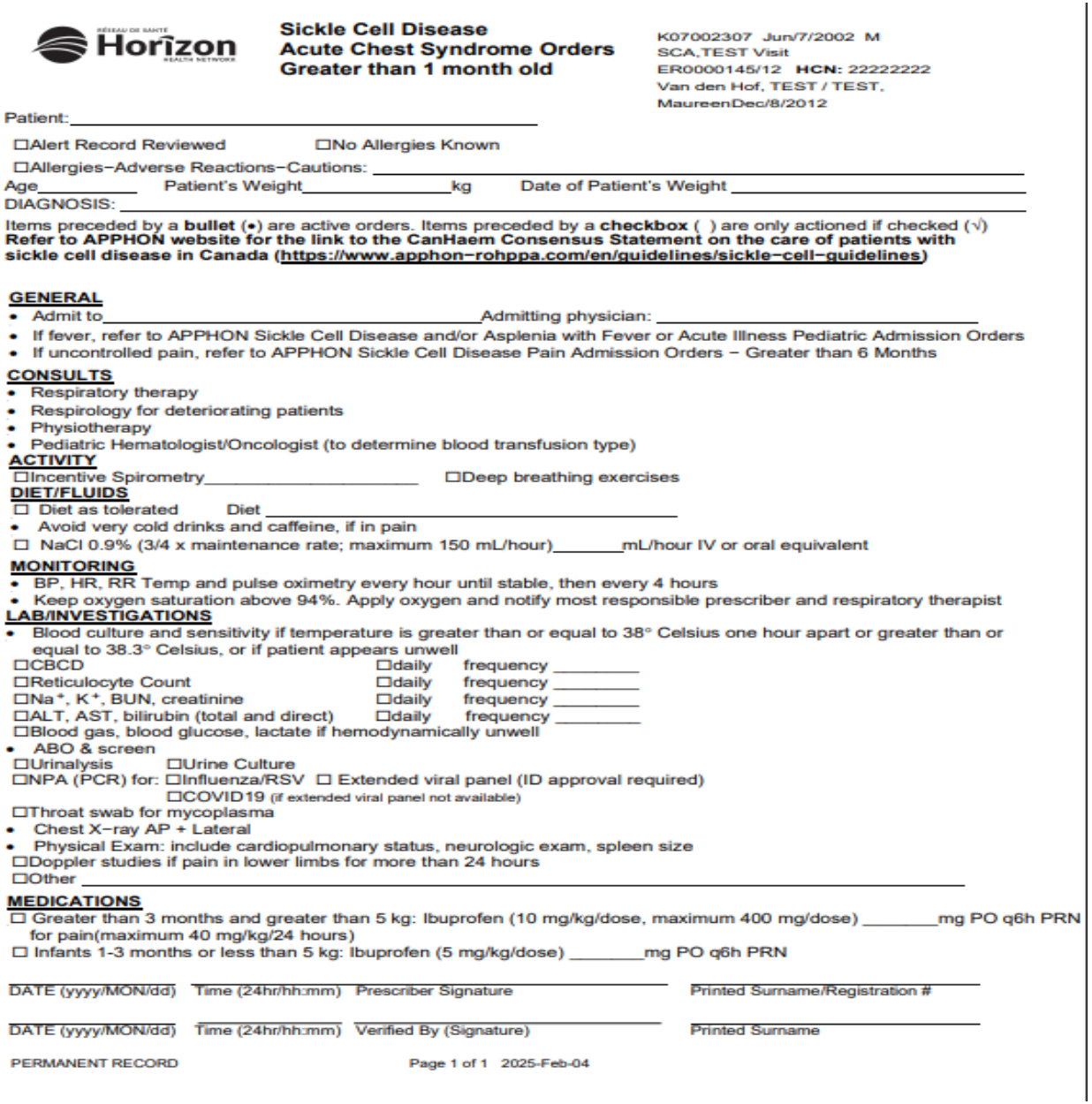
- Vasculature of lungs occluded with Sickled RBC's
- Rapid deterioration of respiratory function
- Second leading cause of hospitalization & primary cause of SCD deaths
- Risk Factors
  - Pain crisis
  - Pneumonia
  - Prior episode of ACS
  - Asthma

ACS Prevention & Treatment

Prevention	Prevention is the key Adequate pain management Avoid over-sedation Incentive spirometry Pulse oximetry Frequent assessment
Management	Transfusion (may include exchange) Pain management Antibiotics Bronchodilators Oxygen



Acute Chest Admission Orders



Horizon

Sickle Cell Disease
Acute Chest Syndrome Orders
Greater than 1 month old

K07002307 Jun/7/2002 M
SCA,TEST Visit
ER0000145/12 **HCN:** 22222222
Van den Hof, TEST / TEST,
MaureenDec/8/2012

Patient:_____

☐Alert Record Reviewed ☐No Allergies Known

☐Allergies–Adverse Reactions–Cautions: _____

Age_____ Patient's Weight_____kg Date of Patient's Weight _____

DIAGNOSIS: _____

Items preceded by a **bullet** (•) are active orders. Items preceded by a **checkbox** ( ) are only actioned if checked (√)
Refer to APPHON website for the link to the CanHaem Consensus Statement on the care of patients with sickle cell disease in Canada (https://www.apphon-rohppa.com/en/guidelines/sickle-cell-guidelines)

GENERAL
- Admit to_____Admitting physician: _____
- If fever, refer to APPHON Sickle Cell Disease and/or Asplenia with Fever or Acute Illness Pediatric Admission Orders
- If uncontrolled pain, refer to APPHON Sickle Cell Disease Pain Admission Orders – Greater than 6 Months

CONSULTS
- Respiratory therapy
- Respirology for deteriorating patients
- Physiotherapy
- Pediatric Hematologist/Oncologist (to determine blood transfusion type)

ACTIVITY
☐Incentive Spirometry_____ ☐Deep breathing exercises

DIET/FLUIDS
☐ Diet as tolerated Diet _____
- Avoid very cold drinks and caffeine, if in pain

☐ NaCl 0.9% (3/4 x maintenance rate; maximum 150 mL/hour)_____mL/hour IV or oral equivalent

MONITORING
- BP, HR, RR Temp and pulse oximetry every hour until stable, then every 4 hours
- Keep oxygen saturation above 94%. Apply oxygen and notify most responsible prescriber and respiratory therapist

LAB/INVESTIGATIONS
- Blood culture and sensitivity if temperature is greater than or equal to 38° Celsius one hour apart or greater than or equal to 38.3° Celsius, or if patient appears unwell

☐CBCD ☐daily frequency _____
☐Reticulocyte Count ☐daily frequency _____
☐Na^+, K^+, BUN, creatinine ☐daily frequency _____
☐ALT, AST, bilirubin (total and direct) ☐daily frequency _____
☐Blood gas, blood glucose, lactate if hemodynamically unwell

- ABO & screen

☐Urinalysis ☐Urine Culture
☐NPA (PCR) for: ☐Influenza/RSV ☐ Extended viral panel (ID approval required)
☐COVID19 (if extended viral panel not available)
☐Throat swab for mycoplasma

- Chest X-ray AP + Lateral
- Physical Exam: include cardiopulmonary status, neurologic exam, spleen size

☐Doppler studies if pain in lower limbs for more than 24 hours
☐Other _____

MEDICATIONS
☐ Greater than 3 months and greater than 5 kg: Ibuprofen (10 mg/kg/dose, maximum 400 mg/dose) _____mg PO q6h PRN for pain(maximum 40 mg/kg/24 hours)
☐ Infants 1-3 months or less than 5 kg: Ibuprofen (5 mg/kg/dose) _____mg PO q6h PRN

DATE (yyyy/MON/dd) Time (24hr/hh:mm) Prescriber Signature Printed Surname/Registration #

DATE (yyyy/MON/dd) Time (24hr/hh:mm) Verified By (Signature) Printed Surname

PERMANENT RECORD Page 1 of 1 2025-Feb-04

Barriers to care for sickle cell patients

1. Lack of knowledge and awareness among health care provides including limited training, negative attitudes, and lack of specialized centers.
2. Systemic Racism and Discrimination including racialized stereotypes, delayed treatment and unequal access to care
3. Socioeconomic Factors including lack of insurance coverage, financial strain and limited access to transportation.
4. Other barriers including lack of standardized care, patient behavior and the stigma associated with sickle cell disease.

Resources

- CanHaem-The Canadian Haemoglobinopathy Association

  Sickle Cell Disease Consensus Statement | Canhaem

- SCAGO- Sickle Cell Awareness Group of Ontario

  Healthcare Professionals Education Program

- Hemoglobinopathy nurse coordinator

Chantale Deyoung
902 470 5085
chantale.riviere@iwk.nshealth.ca

References

- Sickle Cell Disease Canada Website https://sicklecelldiseasecanada.com/
- Atlantic Province Pediatric Hematology Oncology Network website https://www.apphon-rohppa.com/en/guidelines/sickle-cell-asplenia-guidelines
- Association of Pediatric Hematology Oncology Nurses-Hematologic diseases of Childhood 2023