# Optimizing Sickle Cell Disease Care from the Emergency Department to the Inpatient Ward




**Andrea Cervi, MSc, MD, FRCPC**
Hematologist, Windsor Regional Hospital
Physician Lead, Adult Hemoglobinopathy Program
Thrombosis Division Head
Assistant Professor, Division of Hematology
Schulich School of Medicine & Dentistry
Windsor, Ontario, Canada

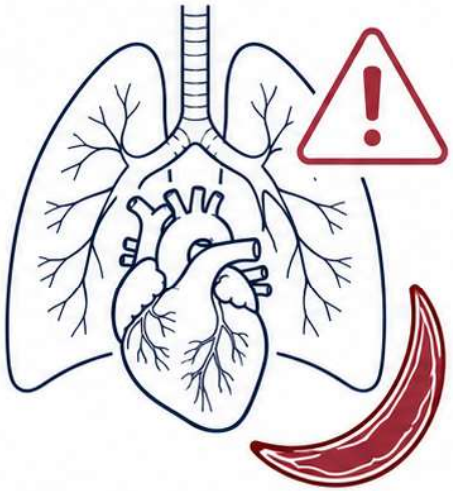
# Learning Objective

By the end of this session, participants will be able to:

## 01

### RECOGNIZE

Common acute complications of sickle cell disease presenting in emergency and inpatient settings



## 02

### APPLY

Evidence-based approaches to timely clinical assessment



**EMERGENCY DEPARTMENT** → **ASSESSMENT & STABILIZATION** → **INPATIENT WARD**

# Speaker Conflict of Interest Disclosure

Dr. Andrea Cervi

## Disclosed relationship

Honoraria received from:

- Novo Nordisk
- Leo Pharmaceuticals

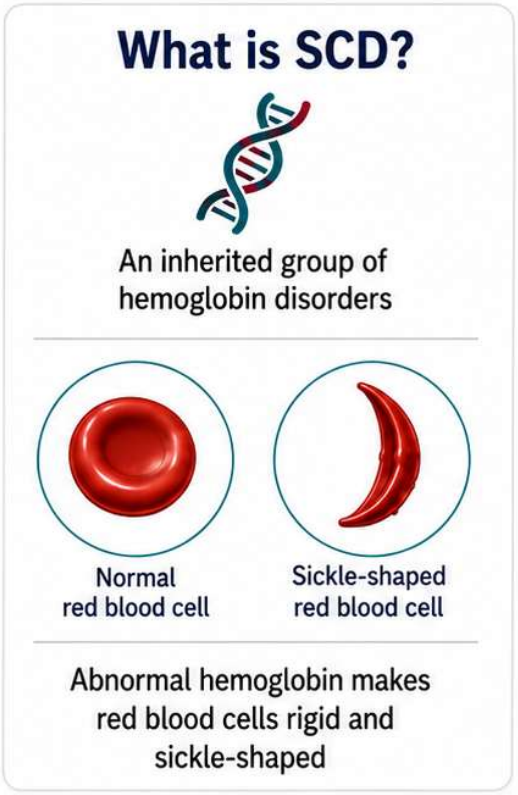
## No other relevant relationships

| | |
|---|---|
| ✓ Advisory boards or speakers' bureaus | None |
| ✓ Funded grants or clinical trials | None |
| ✓ Patents involving a drug, product, or device | None |
| ✓ Other investments or relationships | None |

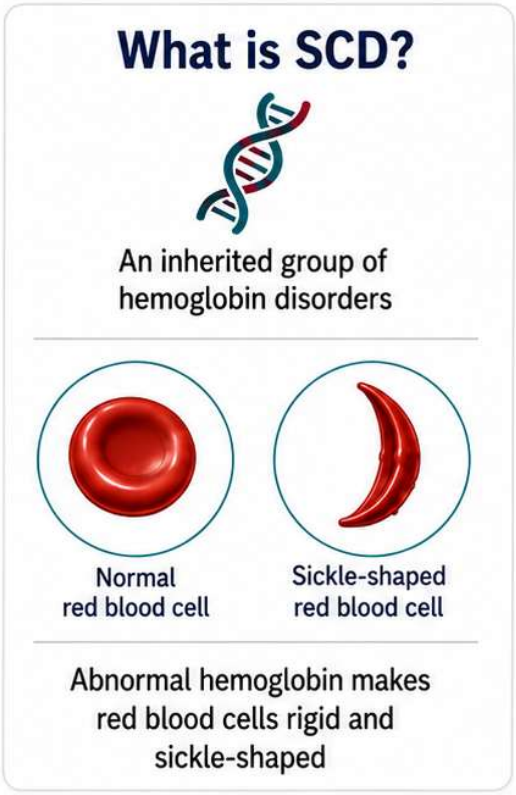
Schulich MEDICINE & DENTISTRY

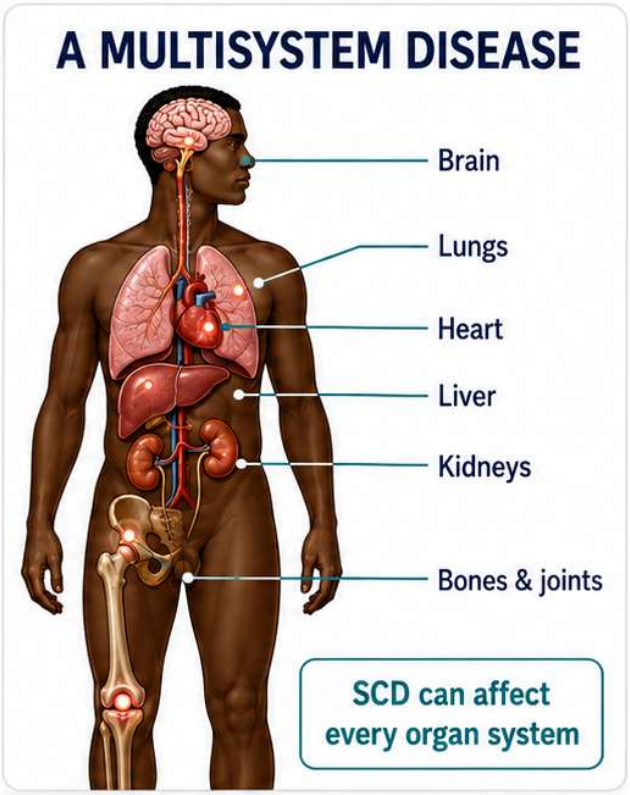
Western

# An Overview of Sickle Cell Disease (SCD)



# An Overview of Sickle Cell Disease (SCD)



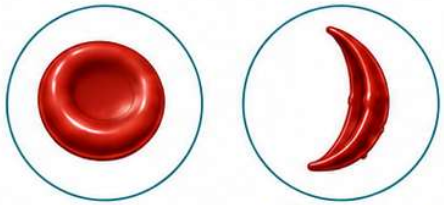



# An Overview of Sickle Cell Disease (SCD)

## What is SCD?



An inherited group of hemoglobin disorders

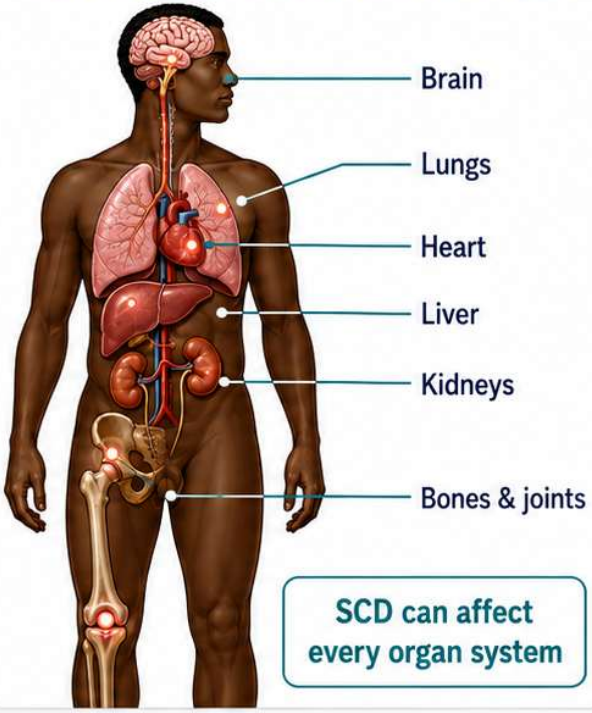


Normal red blood cell

Sickle-shaped red blood cell

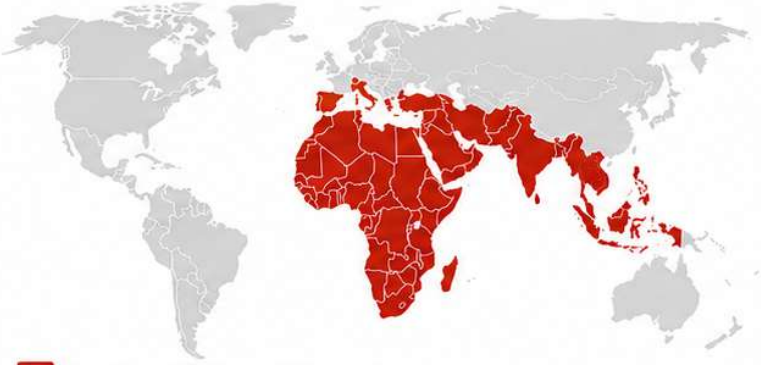
Abnormal hemoglobin makes red blood cells rigid and sickle-shaped

## A MULTISYSTEM DISEASE




SCD can affect every organ system

## Global & Canadian context



Regions with higher SCD prevalence

More than 8 million people worldwide live with SCD



≈3,500 people living with SCD in Ontario

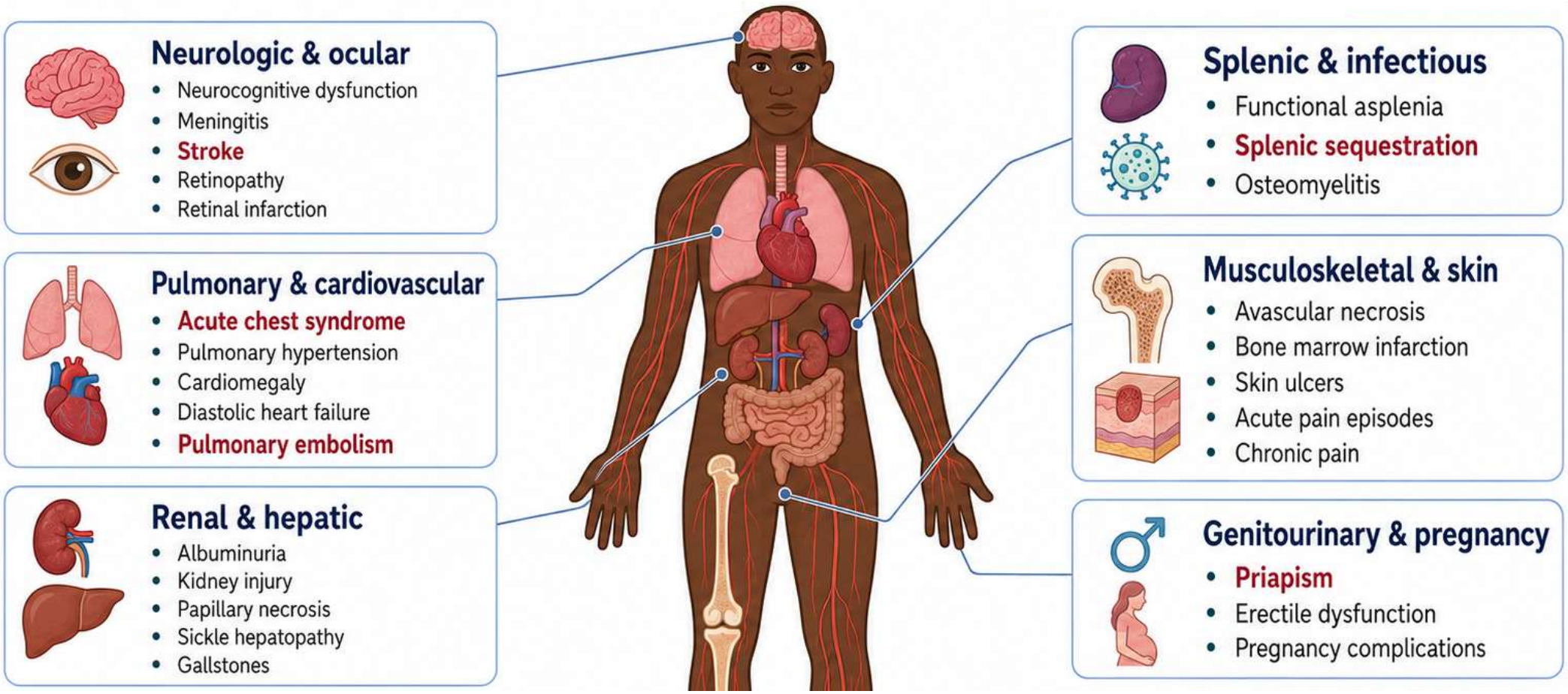


≈6,500 people living with SCD across Canada

**Canada's SCD population is growing. In Ontario, SCD disproportionately affects racialized communities.**

**Sources:** NHLBI Sickle Cell Disease Milestones; Pendergrast et al., CMAJ Open (2023): E724–E733.

# SCD: A Complex Multisystem Disease



# SCD Is Associated With …



**Pain**

**Acute + chronic**

Hallmark vaso-occlusive episodes and chronic pain

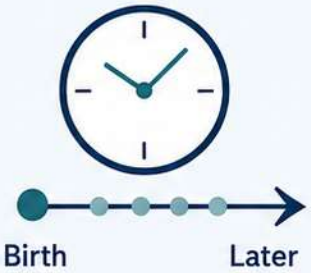
# SCD Is Associated With …



**Pain**

## Acute + chronic

Hallmark vaso-occlusive episodes and chronic pain




**Reduced life expectancy**

## ~20 years shorter

Median difference versus a matched non-SCD cohort

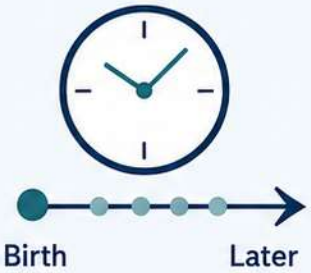
# SCD Is Associated With …



**Pain**

**Acute + chronic**

Hallmark vaso-occlusive episodes and chronic pain




**Reduced life expectancy**

**~20 years shorter**

Median difference versus a matched non-SCD cohort



**Quality of life**

**Substantial impairment**

Scores comparable with those reported during active cancer therapy

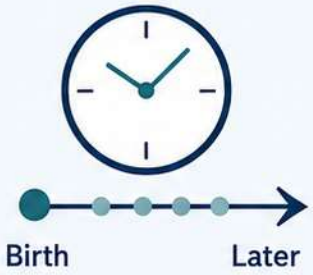
# SCD Is Associated With …



## Pain

**Acute + chronic**

Hallmark vaso-occlusive episodes and chronic pain




## Reduced life expectancy

**~20 years shorter**

Median difference versus a matched non-SCD cohort



## Quality of life

**Substantial impairment**

Scores comparable with those reported during active cancer therapy



## Lost lifetime income

**>$600,000 USD**

Estimated lifetime income loss reported in a U.S. cohort

**SCD affects health, longevity, daily function and economic opportunity.**

Sharma et al. Blood (2007): 5176. • Lubeck et al. JAMA Netw Open (2019): e1915374. • Jiao et al. Blood Adv (2023): 3276–3283.



# 22-year-old male
# HbSS disease

## PAST MEDICAL HISTORY

- Diagnosed with SCD at age 2 years
- Recurrent admissions for vaso-occlusive pain episodes
- Priapism

## HOME MEDICATION

- Hydroxyurea

## SOCIAL HISTORY

- Lives with roommates
- Studying Political Sciences
- From Nigeria; came to Canada 5 years ago

## ED PRESENTATION

Presents to the Emergency Department with a 3-day history of severe pain in the legs and left arm.

> *"This is my sickle cell pain.*

Took three morphine 5 mg PRN tablets at home with no relief.

| | | | |
|---|---|---|---|
| 36.2 °C | **HR** 98 bpm | **BP** 115/76 mmHg | **$SpO_2$** 98% on room air |



# 22-year-old male
# HbSS disease

## PAST MEDICAL HISTORY

- Diagnosed with SCD at age 2 years
- Recurrent admissions for vaso-occlusive pain episodes
- Priapism

## HOME MEDICATION

- Hydroxyurea

## SOCIAL HISTORY

- Lives with roommates
- Studying Political Sciences
- From Nigeria; came to Canada 5 years ago

## CHIEF COMPLAINT

3-day history of severe pain in the legs and left arm.

## ED MANAGEMENT

1. Supplemental oxygen
2. IV hydration
3. Morphine 5 mg PO × 1 dose
4. Persistent pain → referred to Hospitalist for inpatient management



A patient perspective

# “My relationship with the emergency room is that there is no relationship.”

Repeated encounters without trust or continuity can deepen inequity and delay care.

CMAJ CMAJ. 2020;192(31):E900. doi:10.1503/cmaj.201700

pain are listed here.

### Arabs/Muslims

- May not request pain medicine but instead thank Allah for pain if it is the result of a healing medical procedure.
- Pain is considered a test of faith. Therefore Muslim clients must endure pain as a sign of faith in return for forgiveness and mercy. However, Muslims must seek pain relief when necessary because needless pain and suffering are frowned upon.
- Arabs and Muslims prefer to be with family when in pain and may express pain more freely around family.

### Asians

- Chinese clients may not ask for medication because they do not want to take the nurse away from a more important task.
- Clients from Asian cultures often value stoicism as a response to pain. A client who complains openly about pain is thought to have poor social skills.
- Filipino clients may not take pain medication because they view pain as being the will of God.
- Indians who follow Hindu practices believe that pain must be endured in preparation for a better life in the next cycle.

### Blacks

- Blacks often report higher pain intensity than other cultures.
- They believe suffering and pain are inevitable.

### Jews

- Jews may be vocal and demanding of assistance.
- They believe that pain must be shared and validated by others.

### Hispanics

- Hispanics may believe that pain is a form of punishment and that suffering must be endured if they are to enter heaven.
- They vary widely in their expression of pain: Some are stoic and some are expressive.
- Catholic Hispanics may turn to religious practices to help them endure the pain.

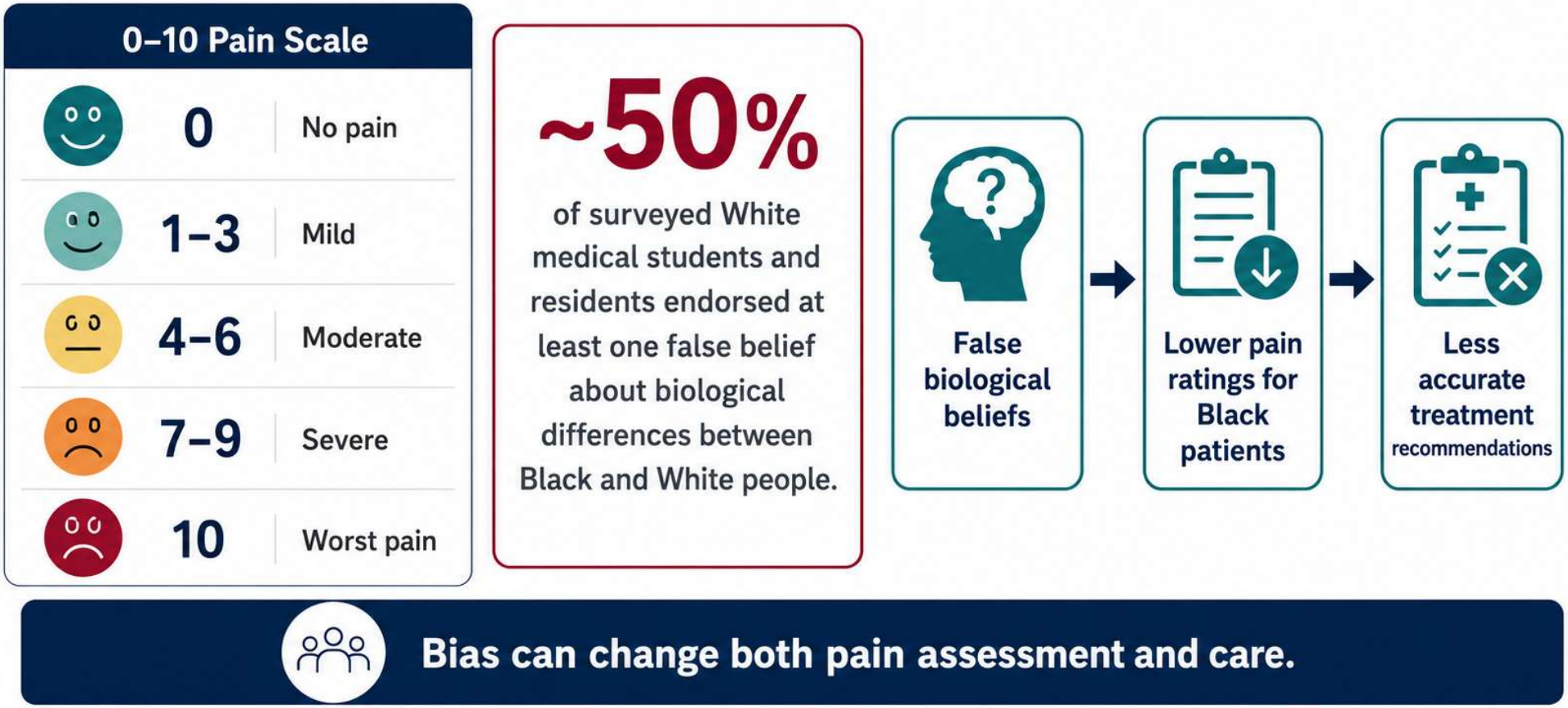
### Native Americans

- Native Americans may prefer to receive medications that have been blessed by a tribal shaman They believe such a blessing allows the client to be more at peace with the creator and makes the medicine stronger.
- They tend to be less expressive both verbally and nonverbally.
- They usually tolerate a high level of pain without requesting pain medication.
- They may pick a sacred number when asked to rate pain on a numerical pain scale.

*Sources:* Based on Munoz, C., & Luckmann, J. (2005). *Transcultural communication in nursing* (2nd ed.). Clifton Park, NY: Delmar Learning; Andrews, M. M., & Boyle, J. S. (2003). *Transcultural concepts in nursing care* (4th ed.). Philadelphia, PA: Lippincott Williams & Wilkins; Al-Atiyyat, N. M. H. (2009). Cultural diversity and cancer pain. *Journal of Hospice and Palliative Nursing, 11*(3), 154–164; Davidhizar, R., & Giger, J. N. (2004). A review of the literature on care of clients in pain who are culturally diverse. *International Nursing Review, 51*(1), 47–55.

*Nursing: A concept-based approach to learning*. Pearson.

# Racial Bias in Pain Assessment




Hoffman KM et al. *Proc Natl Acad Sci USA*. 2016;113(16):4296–4301.

# Ontario Health Sickle Cell Disease Quality Standard

Eight statements supporting equitable, comprehensive care

## Sickle Cell Disease Quality Standard



Ontario Health

# Ontario Health Sickle Cell Disease Quality Standard

Eight statements supporting equitable, comprehensive care

1



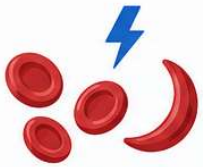
Racism & Anti-Black Racism

2



Comprehensive Health Assessment

3



Vaso-occlusive Acute Pain Episodes

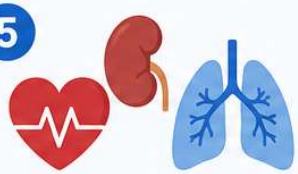
TIME-CRITICAL

4



Life-Threatening Acute Complications

5



Chronic Complications

6



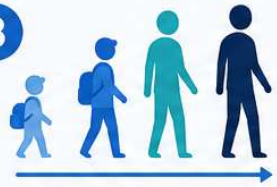
Referral to Health Care Professionals with Chronic Pain Expertise

7



Psychosocial Assessment

8



Transition from Youth to Adult Health Care Services



## QUALITY STATEMENT 3

**Treatment begins within 30 minutes of triage or within 60 minutes of presentation.**

Timely pain and clinical assessment • Shared discharge plan • Follow-up care and support

Source: Ontario Health, Sickle Cell Disease Quality Standard

# Ontario Health Sickle Cell Disease Quality Standard

Eight statements supporting equitable, comprehensive care

1



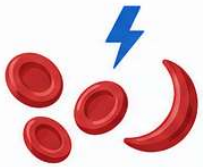
Racism & Anti-Black Racism

2



Comprehensive Health Assessment

3



Vaso-occlusive Acute Pain Episodes

TIME-CRITICAL

4



Life-Threatening Acute Complications

5



Chronic Complications

6



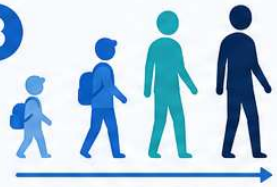
Referral to Health Care Professionals with Chronic Pain Expertise

7



Psychosocial Assessment

8



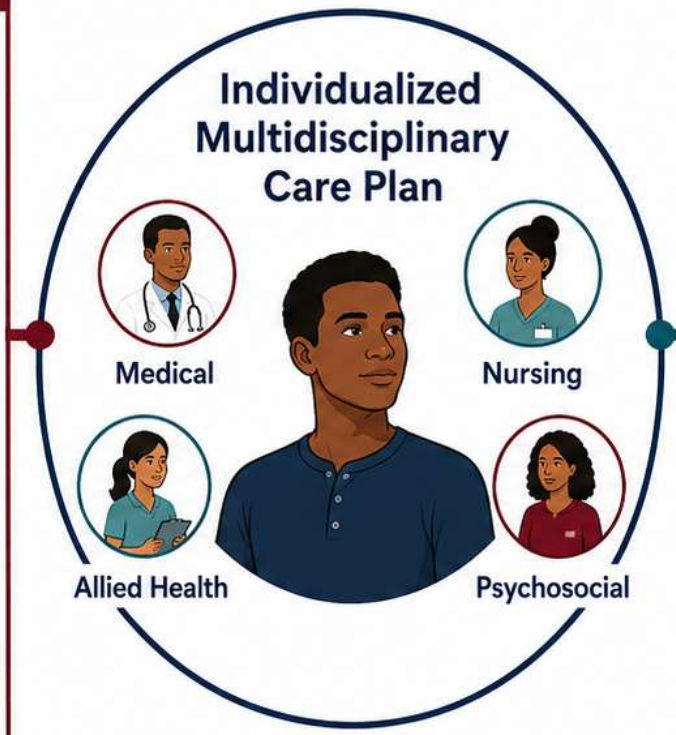
Transition from Youth to Adult Health Care Services

**Prompt, attentive and unbiased pain management is essential.**

The patient is the expert in their own pain.

# Multimodal Pain Management in SCD

## Pharmacologic




## Non-Pharmacologic

# Multimodal Pain Management in SCD

## Pharmacologic



**Opioid Therapy**

Rapid administration with frequent reassessment




## Non-Pharmacologic

# Intravenous Opioid Delivery Options

Goal: rapid, individualized analgesia based on pain severity and prior opioid exposure

## Patient-Controlled Analgesia

PCA

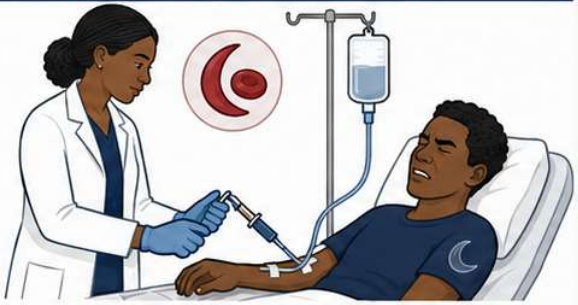


- Patient self-administers small opioid doses
- Programmed lockout interval

Morphine | Hydromorphone | Fentanyl

Supports individualized control during acute vaso-occlusive pain

## Intermittent Clinician Bolus

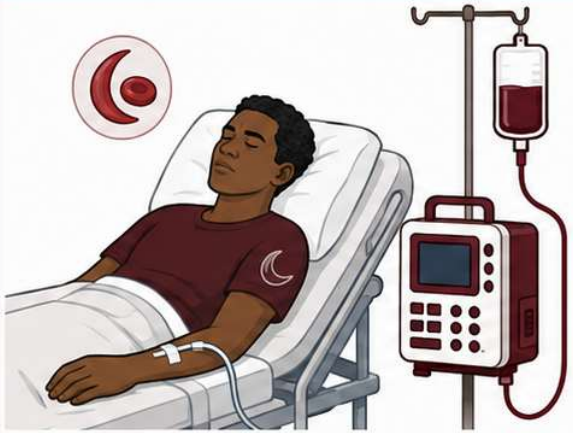


Individualize dosing according to:

- Prior opioid use
- Severity of pain
- Comorbidities

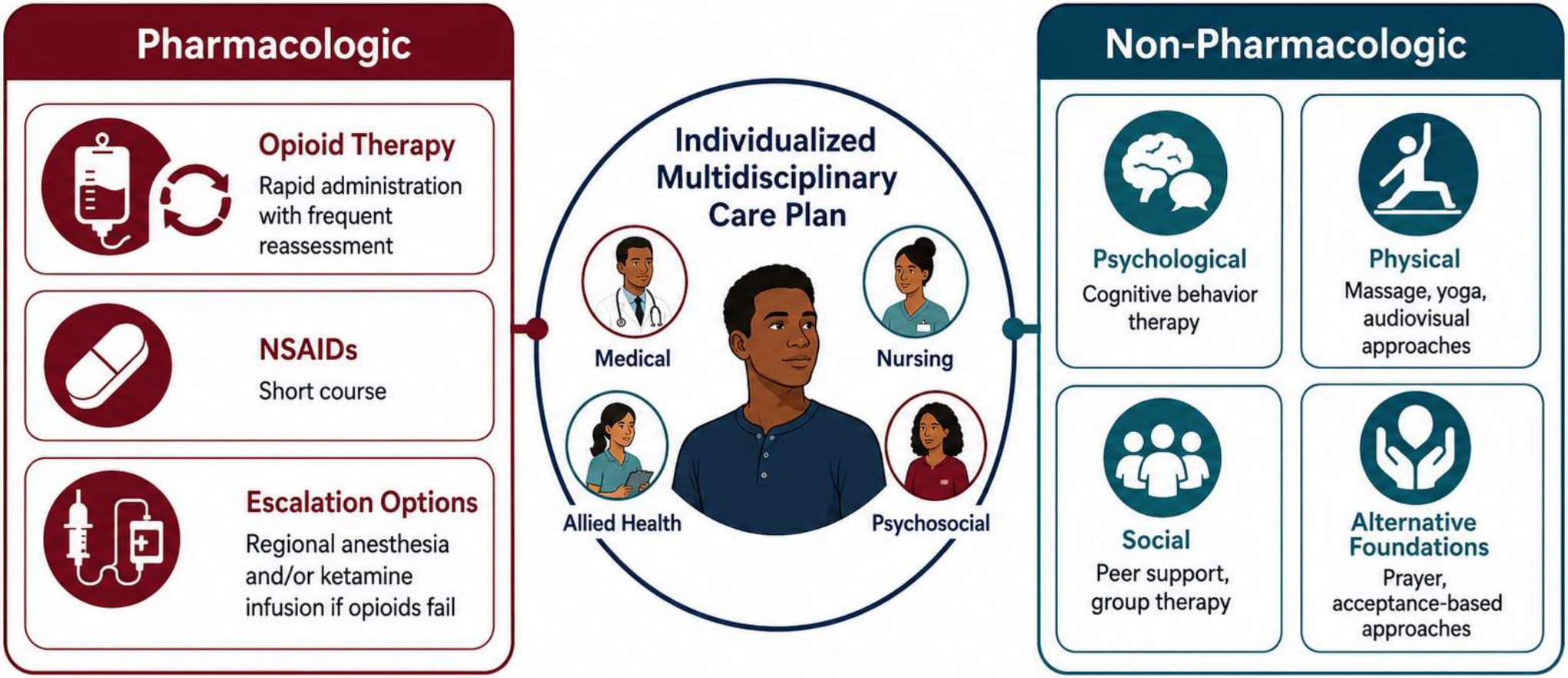
Account for the patient's established analgesic requirements

## Continuous Infusion



Most often used in palliative care and/or ICU settings

# Multimodal Pain Management in SCD



# Adjunct Care Essentials

## Manage Treatment-Related Adverse Effects

## Prevent Inpatient Complications



### Continue Disease-Modifying Therapy

Continue home medications, including hydroxyurea

# Adjunct Care Essentials

## Manage Treatment-Related Adverse Effects



### Constipation

Start a bowel regimen to prevent opioid-induced constipation



### Nausea and Vomiting

Use antiemetics for pain- and medication-associated symptoms



### Pruritus

Use second-generation antihistamines for opioid-induced pruritus

Cetirizine | Loratadine

## Prevent Inpatient Complications



### Continue Disease-Modifying Therapy

Continue home medications, including hydroxyurea

# Adjunct Care Essentials

## Manage Treatment-Related Adverse Effects



### Constipation

Start a bowel regimen to prevent opioid-induced constipation



### Nausea and Vomiting

Use antiemetics for pain- and medication-associated symptoms



### Pruritus

Use second-generation antihistamines for opioid-induced pruritus

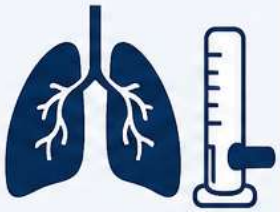
Cetirizine | Loratadine

## Prevent Inpatient Complications



### Fluid Balance

Frequently reassess intravenous fluids to avoid volume overload



### Acute Chest Syndrome

Use incentive spirometry for prevention



### Venous Thrombosis

Provide pharmacologic thromboprophylaxis



### Continue Disease-Modifying Therapy

Continue home medications, including hydroxyurea

RETURN TO THE CASE

# Back to Mr. AM



22-year-old male
HbSS disease

## Relevant Medical History

- Diagnosed with SCD at age 2 years
- Recurrent admissions for vaso-occlusive pain episodes
- Priapism

## Home Medication

Hydroxyurea

## Social History

- Lives with roommates
- Studying Political Sciences
- From Nigeria; came to Canada 5 years ago



## Hospital Course

Admitted with an acute vaso-occlusive pain episode and started on patient-controlled analgesia (PCA)

**On day 2 of admission...**

RETURN TO THE CASE

# Back to Mr. AM



22-year-old male
HbSS disease

## Relevant Medical History

- Diagnosed with SCD at age 2 years
- Recurrent admissions for vaso-occlusive pain episodes
- Priapism

## Home Medication

Hydroxyurea

## Social History

- Lives with roommates
- Studying Political Sciences
- From Nigeria; came to Canada 5 years ago



## Hospital Course

Admitted with an acute vaso-occlusive pain episode and started on patient-controlled analgesia (PCA)

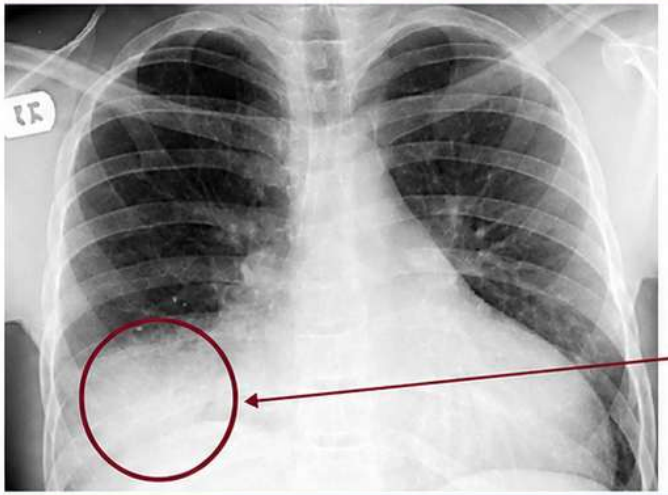
**On day 2 of admission...**

## Clinical Deterioration

| Temperature | Blood pressure | Heart rate | Respiratory rate | $SpO_2$ |
|---|---|---|---|---|
| 39.8°C | 92/40 mmHg | 140 bpm | 40 /min | 95% on 30% $O_2$ by mask |

22-year-old male | HbSS disease | Admitted with an acute vaso-occlusive pain episode

## Chest X-Ray

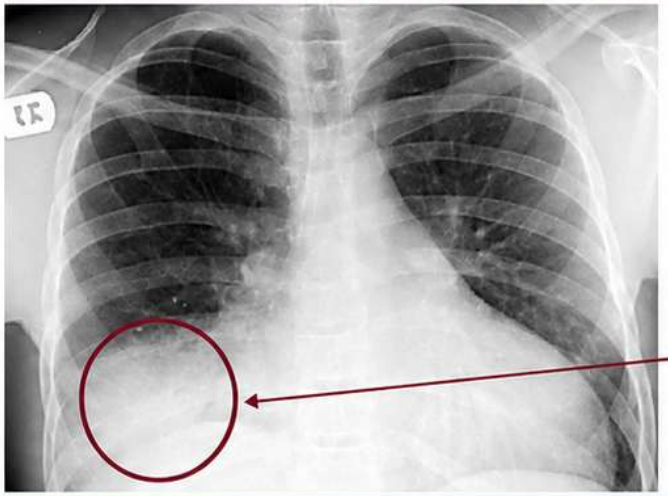



## Clinical Deterioration

- Fever: 39.8 °C
- Respiratory rate: 40/min
- $SpO_2$ 95 % on 30 % $O_2$ by mask
- Blood pressure: 92/40 mmHg
- Heart rate: 140 bpm

22-year-old male | HbSS disease | Admitted with an acute vaso-occlusive pain episode

## Chest X-Ray



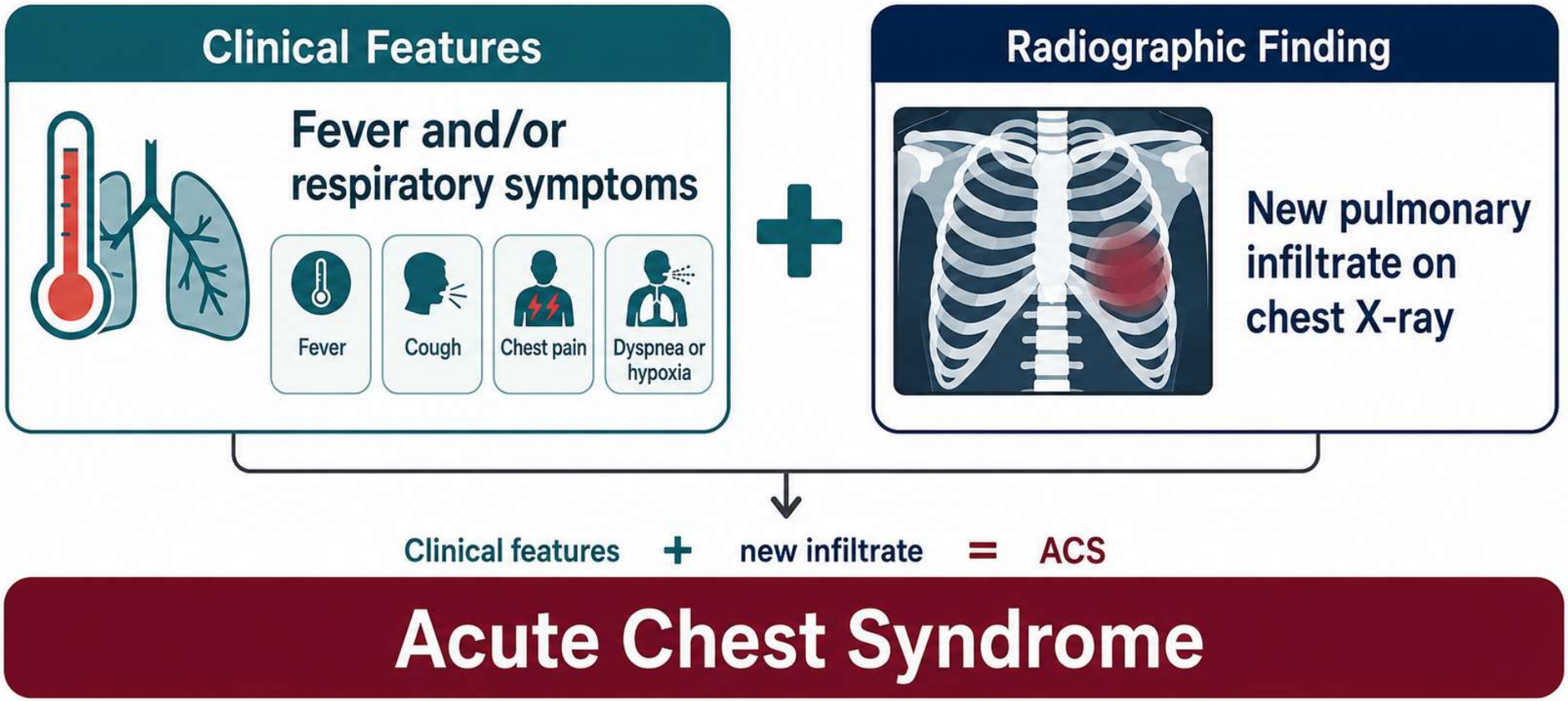

## Clinical Deterioration

- Fever: 39.8 °C
- Respiratory rate: 40/min
- $SpO_2$ 95 % on 30 % $O_2$ by mask
- Blood pressure: 92/40 mmHg
- Heart rate: 140 bpm

Fever + Respiratory symptoms + New pulmonary infiltrate = **Acute Chest Syndrome**

# Diagnosis: Acute Chest Syndrome

# Recognizing Acute Chest Syndrome



# Recognizing Acute Chest Syndrome

## WHAT IT IS

- New pulmonary infiltrate on chest imaging
- Accompanied by fever and/or respiratory symptoms
- A leading cause of hospitalization in SCD

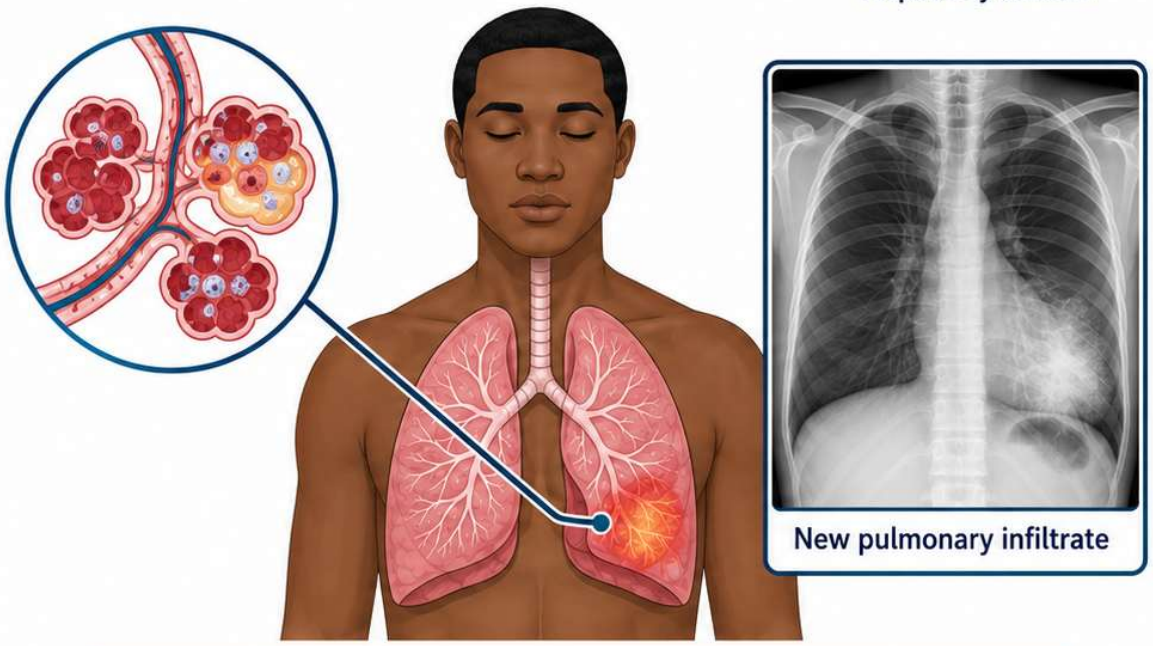
## CLINICAL SPECTRUM

- Severity ranges from mild illness to acute respiratory failure
- May evolve rapidly over hours

**≈50%**

of people with SCD experience at least one ACS episode during their lifetime

Mild respiratory illness → Pulmonary infiltrate → Hypoxemia / respiratory distress



New pulmonary infiltrate

Clinical deterioration can be rapid—monitor closely and reassess frequently.

# Treating Acute Chest Syndrome

## 1 SUPPORTIVE CARE




- Oxygen if hypoxemic
- Judicious IV fluids
- Incentive spirometry

## 2 PAIN MANAGEMENT



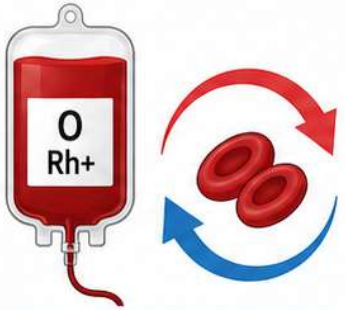
- Individualized multimodal analgesia
- Avoid oversedation and hypoventilation

## 3 EMPIRIC ANTIBIOTICS




- Third-generation cephalosporin
- Plus macrolide / atypical coverage

## 4 TRANSFUSION THERAPY




- Simple transfusion for moderate disease or hemoglobin below baseline
- Exchange transfusion for severe or rapidly progressive ACS

*Avoid hyperviscosity*

## 5 RESPIRATORY ESCALATION




- High-flow oxygen or non-invasive ventilation
- Intubation and mechanical ventilation if needed

# Back to Mr. A.M.

**Acute Chest Syndrome**



## 22-year-old man

HbSS disease



### PRESENTATION

Admitted with an acute vaso-occlusive pain episode complicated by **acute chest syndrome (ACS)**.



### INITIAL BLOODWORK

| | | | | | |
|---|---|---|---|---|---|
| WBC | **16.5** | $\times 10^9$/L | Neutrophils | **12.2** | $\times 10^9$/L |
| Hemoglobin | **65** | g/L | MCV | **78** | fL |
| Platelets | **695** | $\times 10^9$/L | Creatinine | **65** | µmol/L |
| | | | LDH | **800** | U/L |



### INITIAL MANAGEMENT



1. Consult **ICU** for closer monitoring



2. Respiratory therapy assessment; titrate supplemental oxygen to **$SpO_2$ ≥95%**



3. Obtain peripheral blood cultures, urine culture, sputum culture, and viral swab



4. Start empiric **ceftriaxone** and **azithromycin**



5. Group and screen in preparation for **red blood cell transfusion**

# Transfusion in Sickle Cell Disease

## 1. Transfusion targets

| HbS | Total hemoglobin |
|---|---|
| <30% | <110 g/L |

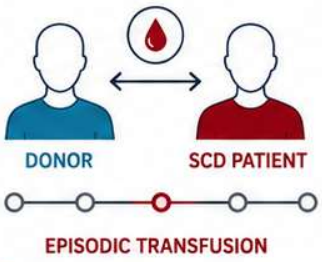
Traditional therapeutic targets

## 2. Alloimmunization risk

≈25%

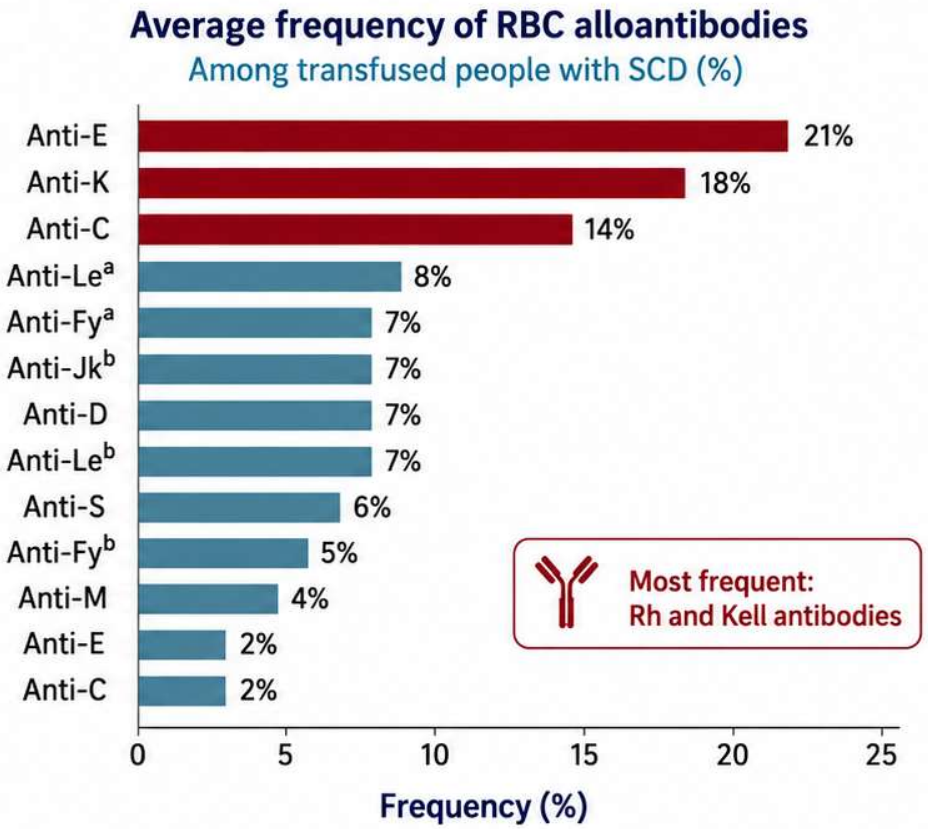
of people with SCD who receive transfusions become alloimmunized

## 3. Why risk varies



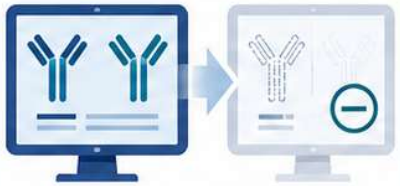

Differences in red-cell antigen expression between donors and people with SCD contribute to risk.

Higher risk with episodic transfusion



# Transfusion in Sickle Cell Disease

## 1 ANTIBODY EVANESCENCE



**>80%**

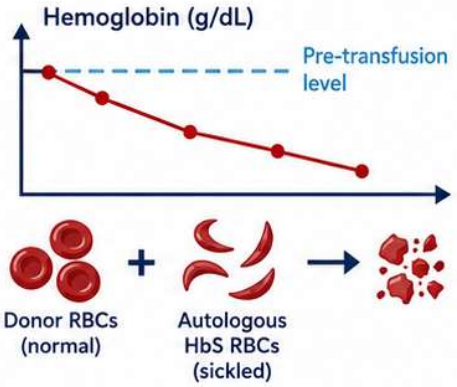
Patients with alloantibodies may later have a negative antibody screen at least once.

## 2 FRAGMENTED TRANSFUSION HISTORY



Episodic transfusions across different hospitals increase the risk of delayed hemolytic transfusion reactions (DHTRs) and hyperhemolysis.

## 3 RECOGNIZE HYPERHEMOLYSIS




Post-transfusion hemoglobin below the pre-transfusion level suggests destruction of both donor RBCs and the patient's own HbS-containing RBCs.

**DHTR + bystander hemolysis**

## 4 PREVENT WITH ANTIGEN MATCHING



Before transfusion, match at minimum for C, E and K; consider extended antigen matching in high-risk patients.

**C · E · K**

**A negative antibody screen does not erase a prior antibody history.**

# Transfusion Modalities

## Simple 'Top-Up' Transfusion

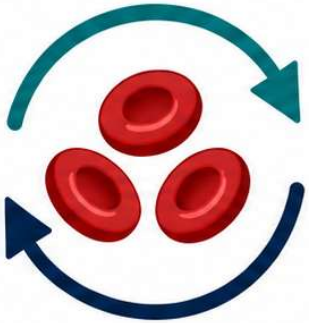


**Hgb ≤90 g/L**

- Use when exchange transfusion is not indicated
- Give 1 unit at a time
- Reassess hemoglobin and symptoms after each unit

**Do not exceed total Hgb 110 g/L**

## Exchange Transfusion



**Hgb S <30%**

- Preferred in acute or high-risk cases requiring rapid reduction of hemoglobin S
- Partial manual exchange or automated apheresis

**Automated apheresis preferred in specialized centres**

# Clinical Indications for Transfusion



## Simple 'Top-Up' Transfusion

- Acute chest syndrome
- Priapism >4 hours refractory to aspiration and irrigation
- Splenic sequestration, intrahepatic cholestasis, or aplastic crisis
- Pre-operative support for moderate- or high-risk surgery



## Exchange Transfusion

- Acute ischemic stroke or retinal artery occlusion
- Acute chest syndrome requiring ventilatory support
- Multi-organ failure
- Fat embolism syndrome
- Stroke prevention in high-risk patients
- Pre-operative optimization for high-risk surgery



Choice of transfusion depends on clinical severity, baseline hemoglobin, HbS target, and hyperviscosity risk.

# Back to Mr. A.M.

**Transfusion Planning**



## 22-year-old man

HbSS disease



### PRESENTATION

Admitted with an acute vaso-occlusive pain episode complicated by **acute chest syndrome (ACS).**



### INITIAL BLOODWORK

| | | | | | |
|---|---|---|---|---|---|
| WBC | 16.5 | ×10⁹/L | Neutrophils | 12.2 | ×10⁹/L |
| Hemoglobin | 65 | g/L | MCV | 78 | fL |
| Platelets | 695 | ×10⁹/L | Creatinine | 65 | µmol/L |
| | | | LDH | 800 | U/L |



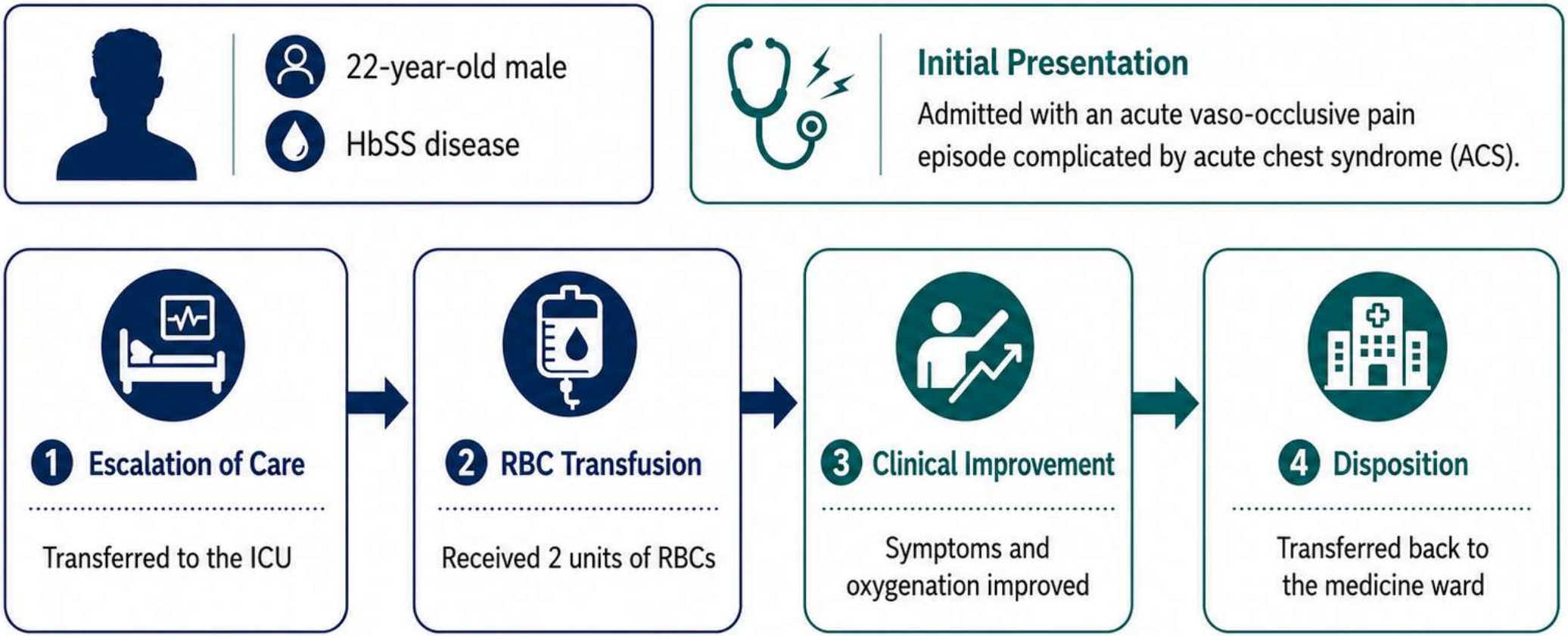
### TRANSFUSION PLAN

1. **Coordinate with the Blood Bank**
   Confirm awareness of SCD and request prophylactic serologic matching for **C, E, and K** antigens at minimum.

2. **Transfuse incrementally**
   Administer **1 unit** of red blood cells at a time.

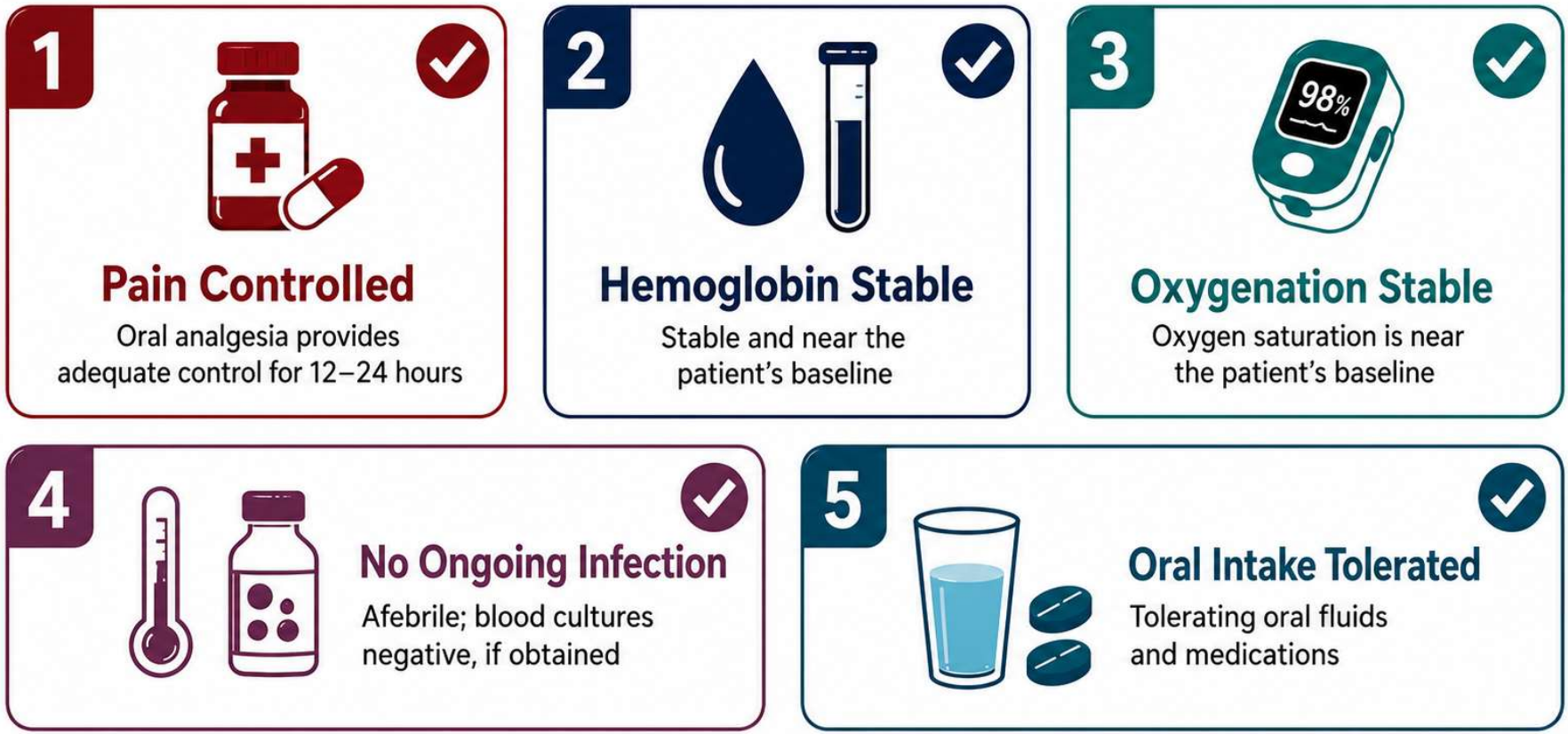
3. **Reassess after each unit**
   Recheck **hemoglobin and clinical symptoms** after each transfusion.

# Back to Mr. A.M.


22-year-old male
HbSS disease
Initial Presentation
Admitted with an acute vaso-occlusive pain episode complicated by acute chest syndrome (ACS).
1 Escalation of Care
Transferred to the ICU
2 RBC Transfusion
Received 2 units of RBCs
3 Clinical Improvement
Symptoms and oxygenation improved
4 Disposition
Transferred back to the medicine ward

# Discharge Planning in SCD

- A comprehensive discharge protocol is crucial once patients are ready to go home



# Patient Education at Discharge

## When to Return to the Emergency Department (ED)

1. Fever: temperature >38°C
2. Progressive pain unresponsive to oral medications
3. Chest pain
4. Shortness of breath
5. Priapism lasting >4 hours
6. New neurologic changes

**Seek urgent assessment**

## Review Home Medications

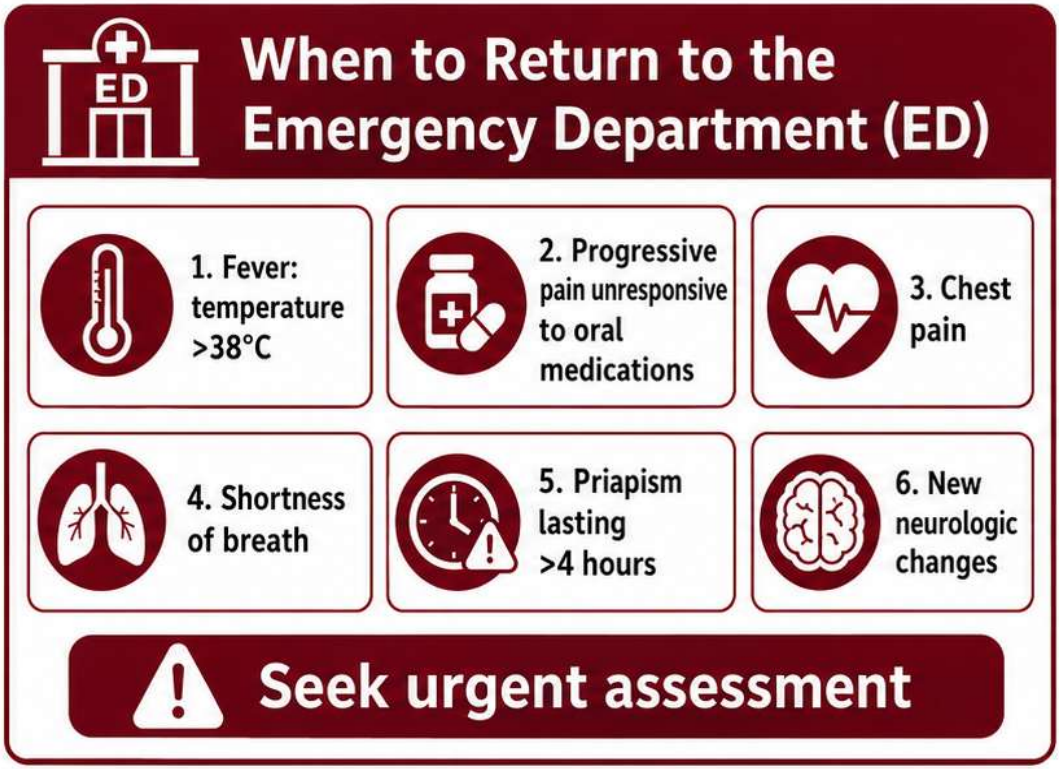
- Continue disease-modifying therapy, including hydroxyurea
- Reinforce medication adherence

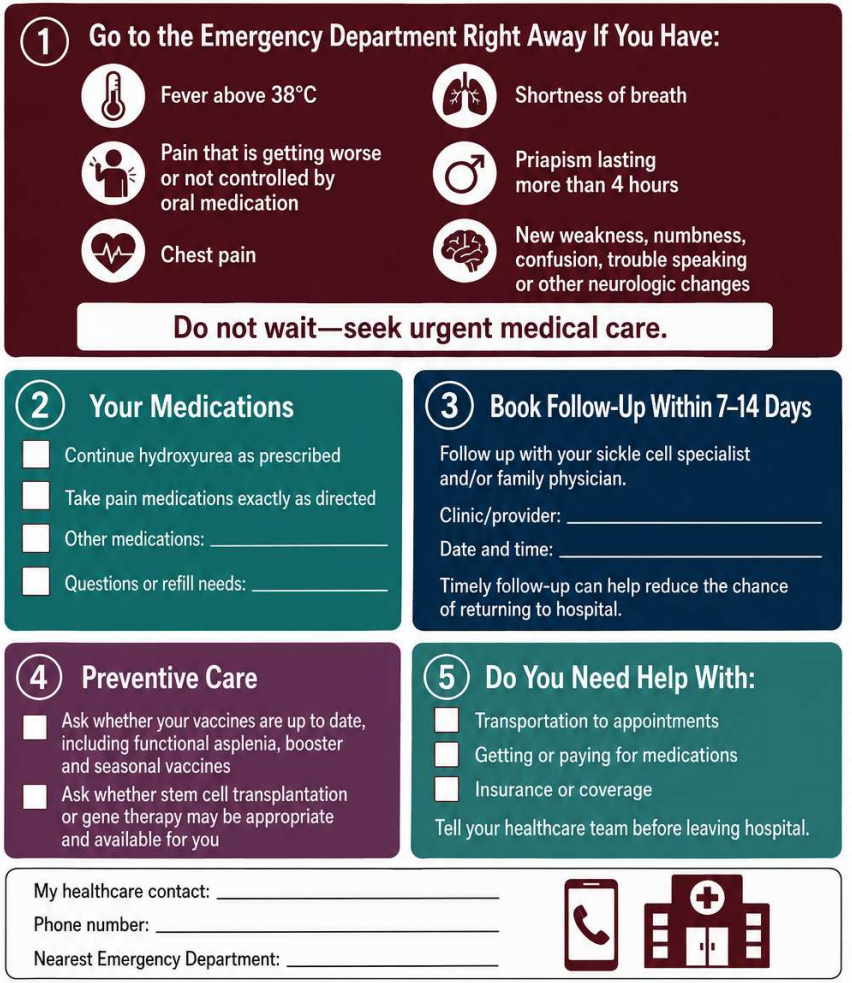
## Confirm Immunizations

Ensure all immunizations are up to date

- Functional asplenia vaccines
- Booster vaccines
- Seasonal vaccines

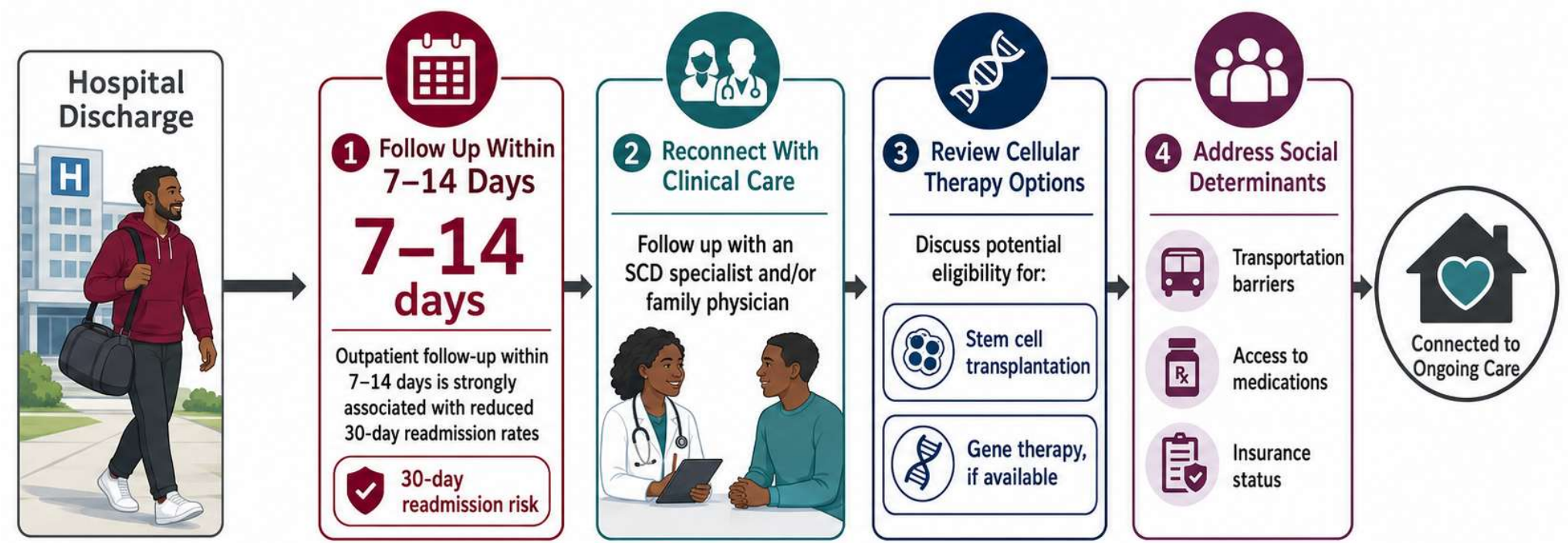
# Patient Education at Discharge




- **Patient facing discharge summary**

# Post-Discharge Follow-Up



# Comprehensive Adult Sickle Cell Disease Care in Windsor

## Windsor Regional Hospital

*Specialized, coordinated care across the adult SCD journey*

### Multidisciplinary Team



**Dr. Andrea Cervi —**
Hematologist



**Lori Clarke —**
Registered Nurse



**Sydney Galloway —**
Social Work



**Julia Thibert —**
Social Work



**Jacqueline Adam —**
Clerk

### Comprehensive Care

Hematology expertise • Dedicated nursing • Psychosocial support • Coordinated follow-up

519-253-5253

wrh.on.ca/SickleCellDiseaseProgram

### Recognized Adult SCD Centre

Ontario SCD Community of Practice

CanHaem
The Canadian Haemoglobinopathy Association

Listed by CanHaem —
The Canadian Haemoglobinopathy Association



Windsor, Ontario

Windsor Regional Hospital

# Optimizing SCD Care Across the Spectrum

## 1 Individualize Acute Pain Care



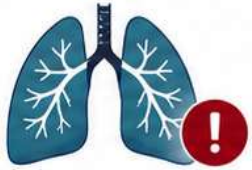

Combine opioids, NSAIDs, and non-pharmacologic strategies with frequent patient input and **frequent reassessment.**

## 2 Prevent Inpatient Complications



Use **incentive spirometry**, thromboprophylaxis, a bowel regimen, and judicious IV fluids.

## 3 Act on Respiratory Symptoms



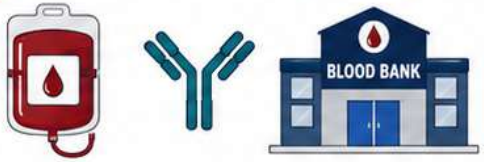
New respiratory symptoms require **urgent assessment** for acute chest syndrome and pulmonary embolism.

## 4 Escalate ACS Management




Provide effective analgesia, broad-spectrum antibiotics, **transfusion when indicated**, and advanced respiratory support.

## 5 Transfuse Thoughtfully




Coordinate with the Blood Bank for **extended red-cell matching** to reduce alloimmunization and hemolytic transfusion reactions.

## 6 Plan a Safe Transition Home

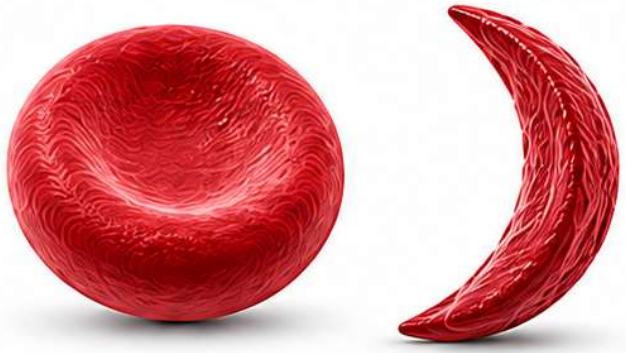


Comprehensive discharge planning and **prompt outpatient follow-up** help reduce repeat hospitalization.

**Patient-centred care** | **Early complication recognition** | **Safe transitions**

# Thank You for Your Attention

## Questions or Discussion?



**Dr. Andrea Cervi**
andrea.cervi@wrh.on.ca