

Clinical Handbook for Sickle Cell Disease Vaso-occlusive Crisis

Provincial Council for Maternal and Child Health &
Ministry of Health and Long-Term Care

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Clinical Handbook: Sickle Cell Disease

Vaso-occlusive Crisis

1.0 Purpose

This Clinical Handbook has been created to serve as a compendium of the evidence-based rationale and clinical consensus for Sickle Cell Crisis.

This document has been prepared for informational purposes only. This document does not mandate health care providers to provide services in accordance with the recommendations included herein. The recommendations included in this document are not intended to take the place of the professional skill and judgment of health care providers.

2.0 Description of Sickle Cell Disease, the Vaso-occlusive Acute Pain Episode and Current State in Ontario

Sickle Cell Disease

Terminology

Sickle cell disease (SCD) causes intermittent and recurrent acute pain episodes as a result of vaso-occlusion. These episodes have been referred to as ‘**vaso-occlusive crises**’ (VOC). **While some have advocated that the term ‘crisis’ be changed, patients still use the terminology. So, for consistency, the term VOC will be used in this handbook.** VOCs are the most common reason for emergency department (ED) visits and hospitalizations for patients with SCD. Ensuring that patients receive timely, effective pain relief is one of the most important elements of delivering high quality care to individuals with SCD in the ED.

SCD is one of the most common genetic disorders. It results in the predominant production of an abnormal/mutant hemoglobin called hemoglobin S (Hb S) in red blood cells. Affected patients usually have at least one S gene inherited from one parent and another abnormal gene S, C, D, E or beta-thalassemia from the other parent. Thus, genotypes of the disease include SS, SC, S-beta thalassemia, SD and SE. Usually, parents who pass on these genes are healthy as they produce more normal hemoglobin called hemoglobin A (Hb A) than abnormal hemoglobins (Hb S, Hb C, Hb D, Hb E). Persons who carry one normal hemoglobin gene and the S gene are known as having sickle trait (AS), and are usually healthy and haematologically normal.

SCD occurs in people of African, Arabic and Indian racial backgrounds. Normally red blood cells are spherical shaped discs. In people with SCD **some of the red blood cells are shaped like a ‘sickle’**, hence the name SCD. The hemoglobin in the red blood cells carries oxygen to all tissues of the body. Sickle shaped red cells, which form when hemoglobin offloads oxygen to the tissues, are more rapidly removed from the circulation by the body (a process called hemolysis) leaving much less hemoglobin circulating (anemia).

The Vaso-occlusive Acute Pain Episode

Normal spherical red cells flow smoothly through the blood vessels while the misshapen (sickled) red cells are sticky and can clump together or stick to the inside of blood vessels causing blockages (vaso-occlusive), in the smaller blood vessels. The blockages result in oxygen deprivation to tissues. The National Health Lung and Blood Institute’s Expert Panel Report on the Management of Sickle Cell Disease describes the vaso-occlusive episode:

“VOCs are typically associated with excruciating pain of sudden onset, some people experience gradual onset of a VOC. Nearly all individuals affected by SCD will experience a VOC during their lifetime. The first VOC may occur as early as 6 months of age, often presenting as dactylitis, but thereafter VOCs occur with variable frequency. VOCs and their accompanying pain most commonly occur in the extremities, chest, and back. When they occur in other sites, they can be

People with sickle cell disease participated in a focus group on July 18, 2015 to share their experiences when they or their loved ones found it necessary to seek care in an emergency department. Throughout this Clinical Handbook the focus group participants’ comments will be documented in text boxes as this note has been.

confused with, or can be the prodromal stage of, other acute complications e.g., head (stroke), flank (papillary necrosis), and abdomen (hepatic or splenic sequestration, constipation from opioid toxicity, or another hepatobiliary complication)). The etiology of the pain must be determined in order to rule out potential causes of pain. A VOC is the hallmark acute complication for persons with SCD and manifests as acute severe pain. VOC can still occur in the presence of other complications. There are no tests to rule in or to rule out a VOC; there are only tests that potentially rule out other causes of pain. Persons with the genotypes HbSS or HbS β^0 -thalassemia are likely to experience more frequent VOCs. Persons with HbAS (commonly referred to as sickle cell trait) do not experience typical VOCs.”

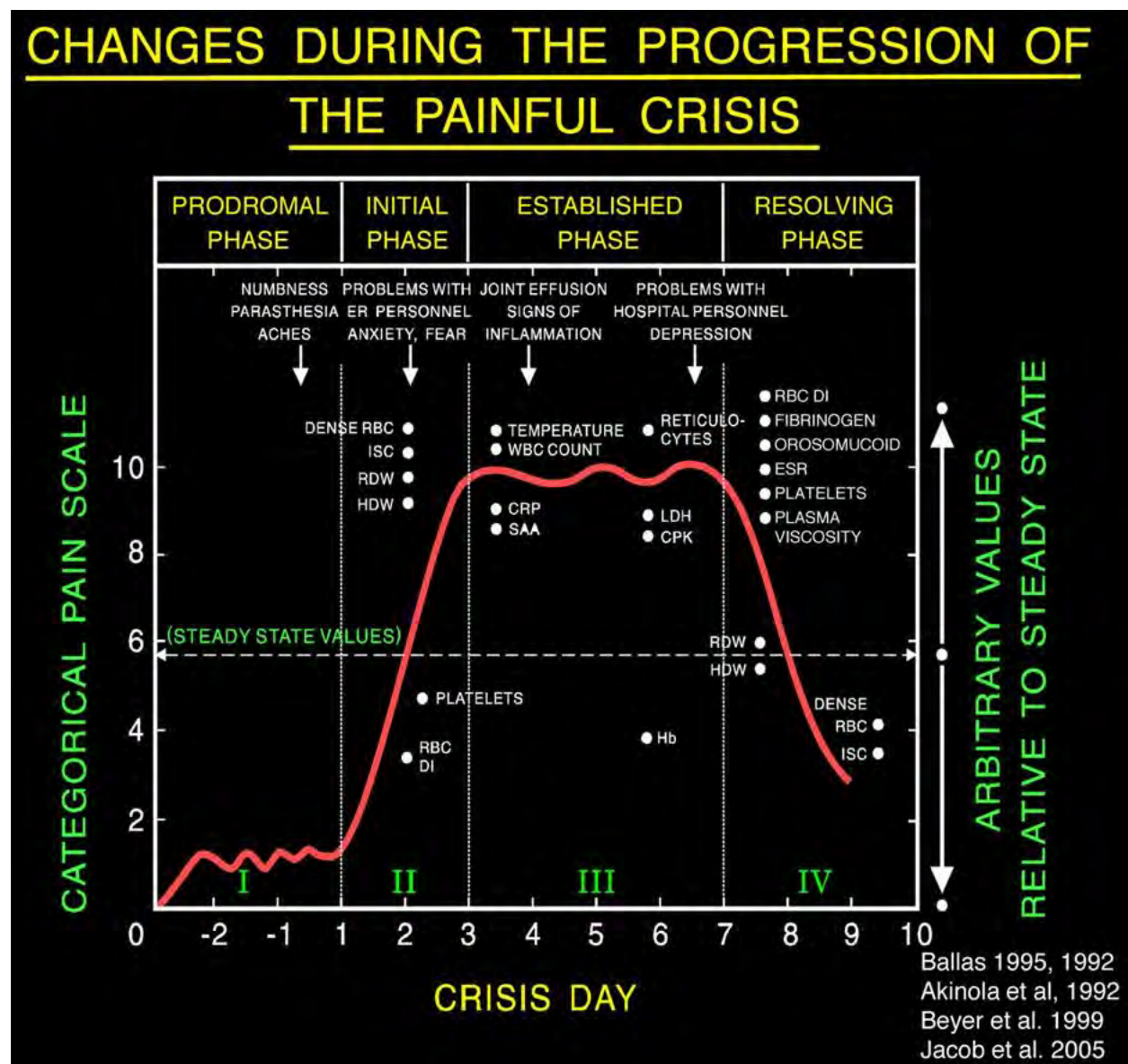
VOC has been described as comparable to or worse than cancer pain, a comparison perhaps arising from bone pain which is associated with some types of cancer and most VOCs. Over time repeated blockages causing hypoxia to the tissues leads to damage to the area, thus SCD becomes a chronic disease with multiple organs and limbs affected to varying degrees. Recurrent episodes of pain, especially if inadequately treated can present further pain management challenges that may require management by chronic pain experts. Thirty percent of people with SCD have some degree of pain 95 % of the time, while fourteen percent have pain less than 5 % of the time^{i, ii}.

They ask you where the pain is on a scale of one to ten. When it is really bad it is so far beyond. It is as if I am being taken ...

Very few disease-modifying therapies exist for use in the treatment of SCD. Hydroxyurea, the most easily available, decreases the frequency of VOCs and reduces organ damage with improved lifespan of people with SCD. It is unknown how many people with SCD are offered or take the medication, or if they or their providers fully understand the benefits. Hydroxyurea does not treat acute pain, rather it reduces the frequency and severity of VOCs. Contraindications to its use are very few. Regular laboratory monitoring of blood is required with the use of hydroxyurea.

Figure 1. Is a schematic of the typical profile of the events that develop during the evolution of a severe SCD VOC in an adult in the absence of overt infection or other complications. Such events are usually treated in the hospital with an average stay of 5-7 days. Pain becomes most severe by day 3 of the crisis and starts decreasing by day 6 or 7.

Figure 1. Typical profile of an SCD VOC.



Current State In Ontario

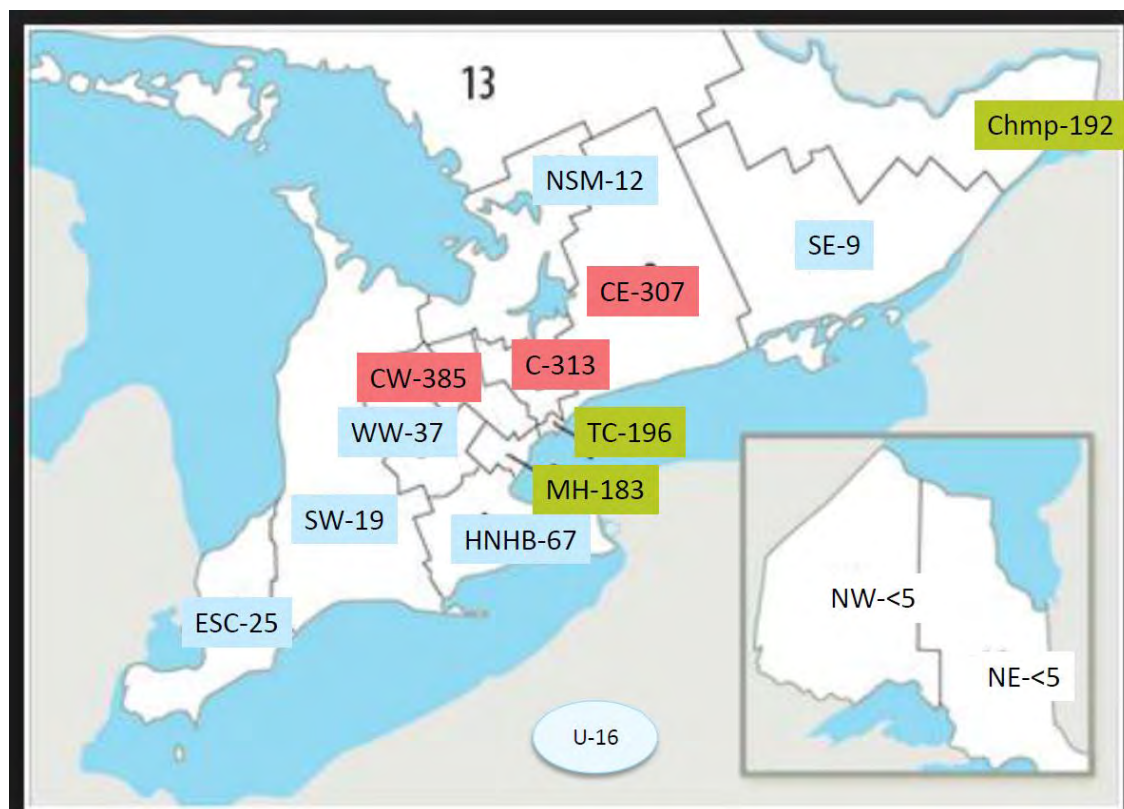
Prevalence Unknown

The number of people living in Ontario who have SCD is not known. Using CIHI data, the number of unique individuals was identified who had SCD and over three years had an encounter, for any reason, with the acute care system and their SCD status documented in NACRS or DAD in the time period from April 01, 2011 and March 31, 2014.

This group is described as follows:

- 59% are aged 18 and over
- 25% are 0 to 9 years
- 55% are female
- Roughly 78% live in the GTA (LHINs 5 to 9)
- 4 communities with over 100 people with SCD
 - Toronto with 698 people (39% of the total patients)
 - Brampton with 270 people (15% of total patients)
 - Ottawa with 179 people (10% of total patients)
 - Mississauga with 163 people (9% of total patients)

Figure 2. Three year acute care encounter volume in Ontario LHINs



U-Unknown LHIN of residence

The distribution of the SCD population is mirrored in the newborn screening data, which demonstrates that the number of people with SCD in Ontario is growing. (Appendix 1) What is not available is the volume contributed by people from other countries who emigrate or have some length of stay in Canada (often for post-secondary education). ED clinicians in paediatric centres and adult centres, particularly in the GTA, Ottawa and Hamilton will encounter people with VOCs. The colour coding of the boxes was simply done to identify common volume ranges by LHIN. LHIN abbreviations: ESC- Erie St Clair; SW-Southwest, WW- Waterloo Wellington; HNHB- Hamilton, Niagara, Haldimand, Brant; CW-Central West; MH- Mississauga Halton; TC-Toronto Central; C-Central; CE-Central East; SE-Southeast; Chmp – Champlain; NSM- North Simcoe Muskoka; NE- North East; NW- North West

Table 1. Ranking Hospitals by Volumes of ED Visits and Admissions for VOC presentations

Hospital	Ranked ED Volumes	Ranked Hospital Admissions
Brampton William Osler	1	1
SickKids	3	2
Toronto General	2	5
Trillium Credit Valley & Mississauga	4	4
Humber River Regional	5	6
Rouge Valley (Toronto)	8	3
The Ottawa Hospital	6	7
Scarborough Hospital	7	9
North York	9	8
CHEO (Ottawa)	12	10
Toronto East General	11	12
Hamilton Health Sciences	12	11
Mount Sinai (Toronto)	10	16

Further detail regarding ED visits / revisits and admissions / readmissions in high volume hospitals is provided in Appendix 2.

Table 2. Average number and per patient encounters to the ED and hospitalizations for VOC episodes for all Ontarians with SCD, by age category

Patient age	Unique individual acute care encounters all reasons(3 year)	ED Visits Yearly average (2011-13/14) (ED visits/encounters)	# Hospitalizations Yearly average (2011-13/14) (Hospitalizations/encounters)
1. 00-09	440	242 (0.55)	200 (0.45)
2. 10-17	288	191 (0.66)	190 (0.66)
3. 18-25	302	430 (1.42)	264 (0.87)
4. 26-39	382	557 (1.46)	268 (0.70)
5. 40+	356	192 (0.54)	82 (0.23)

ED visits and hospital admissions per person increase with age, with 18-39 years olds having the highest average of ED visits / admissions per person. Above the age of 40 years, increased mortality in those with severe disease and better experience with managing and living with pain may account for reduced ED visits and hospitalizations.

CTAS Level 2013/14

The Canadian Emergency Department and Triage Acuity Scale is used by triage nurses to determine in a 2 to 5 minute assessment how quickly a patient needs to be seen by a physician on arrival at the ED. CTAS level is a guide and compliance with adhering to time frames is relative to the CTAS levels of other patients in the ED. The highest CTAS is level 1 and the patient must be seen immediately. Level 2 is for patients who are **considered 'emergent' and the expected time to physician assessment is less than or equal to 15 minutes**; the 15 minute interval also guides the frequency with which a nursing reassessment should occur while awaiting physician assessment if there is a delay. CTAS level 3 is considered **'urgent' and requires that the patient is seen in less than or equal to 30 minutes**.

"When I was a kid my mom used to call my doctor and we would take a cab to the hospital and they would be waiting for me with a bed and IV and pain medication. My first crisis as an adult after I turned 18 was like a slap in the face...it was brutal. It took four hours until they gave me anything."

Adults with SCD are more often assigned the lower CTAS level of 3 compared to those less than 18 years old (Table 3) and even when they are assigned a CTAS level of 2, adults typically wait longer to be seen by a physician and have a longer length of stay in the ED (Table 4).

Table 3. Percent of patients assigned CTAS level 3 of all assigned level 2 or 3 by age category

Fiscal 2013/14	Percent Urgent of (Urgent + Emergent)
Age Group	$L3/(L3+L2)*100$
1. 00-09	13
2. 10-17	9
3. 18-25	28
4. 26-39	26
5. 40+	47

"I am afraid to go to a hospital now. I will not step foot in a hospital."

Table 4. Comparing paediatric and adult CTAS levels and ED wait measures in fiscal 2013/14

Fiscal 2013/14		*Ages 00-17	Ages 18 and up
CTAS Level 2 –Time (in hours) from triage to initial physician assessment	cases*	405	749
	Mean	0.7	1.5
	Median	0.5	1.1
	P90**	1.4	3.1
Time (in hours) from triage to Disposition Decision in ED	cases*	469	1124
	Mean	4.7	7.8
	Median	4.3	6.9
	P90**	7.7	13.8
Time (in hours) from triage to patient leaving the ED	cases*	469	1122
	Mean	6.8	12.1
	Median	6.2	9.3
	P90**	10.7	22.8

"It is a fear of how you will be treated."

"I am afraid I will be sent home when I don't feel well."

*Number of cases does not include those with negative wait times, missing date/time information and cases where the patient left without being seen or died.

**P90 is the time by which 90 percent of patients have been seen.

Barriers to Care

The first measure in table 4 is the time the triage nurse sees the patient (time 0) to the time the physician interview or assessment (PIA) takes place. The many potential enablers and barriers in this aspect of care reflect the complexity of the ED environment:

Tools and processes as enablers and barriers:

- A **medical directive or other “protocol” mechanism** will enable the nurse to initiate an intravenous (IV) ASAP
- Once the physician sees the patient and decides to order opioids, other barriers to quick administration can include CPOE, (computerized physician order entry) dispensing cabinets, limits on dose/rate at which the narcotic can be given, needs of other patients
- Establishing medical directives, pre-printed orders and developing a culture of MD-RN communication regarding pain control can reduce the time to analgesic administration.

Clinical challenges

- IV access can be challenging (children, dark skin, lots of IVs in the past so no easy sites)
- SCD patients with a VOC often do not exhibit painful distress; which is what ED staff respond to when providing early analgesia

“When it is painful it is really painful so you don’t really want to live. I don’t want to go to the hospital. I wish doctors got the urgency to help people.”

“.....frustrated with having a voice that gets drowned out by other conditions”

Nurses’ role

- Once the nurse determines that the patient is likely having a VOC and has asked about symptoms of complications, they may attempt to obtain an order for opioids even prior to physician initial assessment (PIA)

Physicians’ Role

- Some physicians would see a patient experiencing substantial pain and/or note their sickle cell history, begin an assessment with ABCs and then move to pain and find a nurse to administer analgesia.
- Others would complete a full assessment and then leave written orders for analgesia and investigations for the RN to complete.
- Being seen by a junior trainee in a teaching hospital or by an ED physician in a community hospital with little experience managing SCD patients can easily delay treatment further after PIA
- In a community hospital a paediatrician may be the expert consulted for a paediatric patient with VOC.

“He ended up needing a transfusion overnight. There was a nurse there every hour like clockwork. You really felt like he was getting cared for. I’m frightened for when he turns 18.”

Patient - Provider Barriers to Care

- Patients experience negative provider attitudes of distrust, inconsistencies in care and attitude, lack of respect, etc.
- Patients arrive at ED in pain and with the trepidation about whether they will receive timely, adequate and compassionate care.
- The net result can be a confrontational environment.

Patients with High ED / Hospital Utilization

“Individuals with more than three hospitalizations for a VOC in a year are at an increased risk of early death.” (NHLBI 2014)

A small number of people with the disease are the highest users of ED visits. Provincially, over the three fiscal years from 2011/12 to 2013/14, 2.7 percent of patients accounted for 27.5 percent of visits and 17 percent of patients accounted for 53 percent of visits for VOC. (Table 5) As previously stated the disease becomes chronic over time and with unpredictable episodes of acute pain and chronic disease, there are psychological consequences and economic consequences. **A small percentage of patients make up a large percentage of ED visits.**

“You have to speak very calmly and yet be assertive. You have to make sure you are presentable. Even if it is two in the morning and your child is screaming in pain take a moment to brush your hair and dress well. I’ve coached my son on how to speak to nurses and doctors, not to look down but look them in the eye when you speak to them.”

“It is all about strategy. Figuring out what you will say to them and how to say it to get them to help you. It is like a game of chess.”

“One time a young doctor put her hand on my son’s arm and said to him ‘You do not need to manage your mother’s anger’. I was standing right there.”

“It takes a lot to put yourself in the place (mentally) to go to the hospital. It is like a week in a battle zone for minimal release.”

Table 5. High users of ED Among People with SCD experiencing VOC

ED Visits SCD VOC /Fiscal Year	2011	2012	2013	3 year average
Unique patients with ED visits	602	596	627	608
Number of visits	1692	1597	1597	1629
People \geq 4 visits	89 (15%)	100 (17 %)	93 (15 %)	17%
Number of visits by people \geq 4 visits	921 (54%)	871 (55 %)	806 (50 %)	53%
People with \geq 10 visits	15 (2%)	17 (3%)	18 (3%)	2.7%
Number of visits by people \geq 10 visits	525 (26%)	422 (26%)	399 (25%)	27.5%

A systematic review demonstrated that provider knowledge and attitudes toward people with SCD are among the barriers to careⁱⁱⁱ. The waits for care in the various measures in the ED together with the experiences described by the focus group participants suggest that this is generalizable to Ontario.

My brother was 33 years old when he went to ED with a crisis. They saw a black male and told him he was looking for drugs not having a crisis. Despite that comment they gave him 40 dilaudid and sent him home. I called him and told him I would go over the next day as he lived alone in an apartment. My aunt called in the morning and told me not to come. They found him dead. He had a pill sitting on his tongue. When you are in a crisis you don’t know what’s two hours or four hours....”

Table 6. SCD Expert Panel Members Contextualize the Current State Data

Metric	Adults	Paediatrics
Volume of ED visits (as a percentage of all acute care encounters)	<p>ED visits rise in the 18-39 year old group. This is also identified in published literature.</p> <p>Factors thought to include:</p> <ul style="list-style-type: none"> • Increasing severity of disease • Increasing chronic component of disease • More adults receive care outside of tertiary comprehensive programs • Suboptimal transition from paediatric to adult health services 	
CTAS Level and physician interview / assessment (PIA)	<p>CTAS level 3 is more often assigned to adults than children. Greater number of competing priorities in adult ED. Patients may present without an advocate present. Patient response to pain variable; ranges from stoic and not communicative to outraged and calling out for pain medications. Higher percentage of children use ED at tertiary centre medical home, whereas adults use community ED closer to home.</p>	<p>Paediatric patients presenting to ED are most likely to be assigned CTAS level 2.</p> <p>Paediatric patients are accompanied by parents.</p>
Triage to physician interview / assessment (PIA)	<p>The recommended time increment is linked to the CTAS assignment.</p> <p>Even when CTAS level 2 is assigned, adults wait longer for PIA, the time by which 90% (P90) of people are seen is very high for both CTAS level 2 and 3.</p> <p>For this metric, the goal should be that 90 percent of people meet the target. P90 is the first metric to look at to reduce variation, and then to improve overall. Setting goals based on the ideal care is the only way to push improvement.</p>	<p>Children are more likely to be assigned CTAS level 2 and are seen more quickly.</p>
Triage to disposition	<p>The patient needs intensive treatment and monitoring to have a reasonable chance of getting the pain under control. Once the intensive treatment has been provided the decision can be made if the VOC severity is such that the patient feels they can continue with home management or if they need to be admitted. Setting targets for the disposition is consistent with the recommendations of prompt intensive treatment and a decision based on that care having been delivered.</p> <p>Improvement across this measure is needed at all points; P90 mean and median all high.</p>	<p>As with time from triage to PIA, the paediatric metrics are better on this, however there is room for improvement at the P90 measure.</p>
Admitted patients: Discharge disposition to time patient leave ED	<p>If triage to disposition time is good and there is a long wait to leave the ED for patients who are admitted, that is outside of the ED team's control (mostly), as patients often need to wait for a bed.</p>	<p>The same pattern of variation is seen with paediatric measures - better than adult, worse from 10 to 17 than 0 to 9 years old.</p>

Metric	Adults	Paediatrics
Percentage of patients who attend ED that are admitted	Variable across organizations and may not be a valuable metric given the tendency for sicker patients to seek care in tertiary centres and for the high rates of revisits associated with low rates of admission. As knowledge and care processes improve across the system this may be a metric to reconsider. Data currently available does not allow for explanation but highlights variations across institutions.	Higher percentage of paediatric patients are admitted from ED. This may reflect more frequent fever or concern for fever in pediatrics.
Repeat ED visit or inpatient admission ≤ 14 days	<p>This is a key target of the QBP recommendations.</p> <p>Rapid, aggressive treatment, appropriate discharge disposition and adequate discharge preparation / planning are core guidelines to reduce ED revisits and inpatient readmissions.</p> <p>Adequate discharge preparation and planning will include individualized intensive case management for the small group of patients who are frequent users of acute care.</p>	Much lower in paediatric cases and particularly in tertiary and existing satellite centres. A higher percentage of children are linked to comprehensive care centres as medical homes.
Length of inpatient stay	<p>LOS is longer in adults, which may reflect the increasing complexity of the disease and pain presentations with aging and cumulative end-organ damage, although other factors are likely also at play. This has also been reported in the literature.</p> <p>When looking at the LOS, look mostly at the median, which is quite consistent across the board, and is similar for pediatric and adult.</p> <p>P90 LOS likely reflects patients with more complex pain or medical issues.</p>	
Repeat ED visit or inpatient admission ≤ 30 days	<p>The data show that for the bulk of patients who return to ED or are readmitted this occurs within the first 14 days, however there is a group that do so > 14 days and ≤ 30 days. This is believed to be a failure of care and coordination between the hospital and community based providers and / or ambulatory clinics.</p> <p>Again, the small number of high users of acute care is thought to make up a significant portion of this group.</p>	
High users of acute care	<p>It has been identified that use of acute services is higher between the ages of 18 and 39 years. However, it is a small proportion of users who use a high proportion of resources.</p> <p>The chronic care models with intensive case management will be essential to curbing the ongoing high use of acute care. This subset of patients requires additional interventions.</p>	

Table 7. Issues and Gaps cross the Care Path Identified by Expert Panel Members

Patient, Provider and System Issues and Gaps Along the Clinical Path of Person with Sickle Cell having Pain Due to a Vaso Occlusive Crisis							
Themes: Education, Communication, Coordination							
	Presentation/ Location / Seeking help	Triage	Initiation	Monitoring	Decision making: Admission vs Discharge and planning	Follow - up	Preventing unplanned ED, unplanned visits, Repeat visit
Patient	<div>Home based management</div> <div>Patient education</div> <div>Awareness of phone advice options</div> <div>Adherence to care plan / medical advice/ follow-up</div> <div>Past experiences with acute care</div> <div>Self care plans</div> <div>Acceptance of SCD diagnosis</div> <div>Access to meds</div> <div>Availability of transportation / Mode of transportation-public, cars, EMS</div> <div>Social circumstances</div>	<div>Chronic pain and opioid use</div> <div>Subjectivity of pain (Lack of) outward manifestations of pain</div> <div>Patient and family input into care plan</div> <div>Behaviours and interactions with care providers</div> <div>Patients to have own copy of care plan</div>	<div>Challenge getting IV access</div> <div>Socioeconomic status / area</div>	<div>Support person</div>	<div>Does patient feel ready to go home?</div>	<div>Priority on attending routine follow-up</div> <div>Cultural issues</div> <div>Language barriers</div> <div>Access to meds and education about their use</div>	<div>If unplanned visit to ED: (probe) psychosocial circumstances</div> <div>Tools for prevention eg. stress reduction</div> <div>Geographic location</div>
Provider	<div>Knowledge of SCD</div> <div>Education to patients</div> <div>Inquire about opiate supply at routine visits</div> <div>Access to expert advice</div> <div>Availability of providers</div>	<div>Education of triage</div> <div>Hesitation about narcotic doses</div> <div>Stigma: drug seeking</div> <div>Obtain adequate history of opioid use / dose requirement</div> <div>Judgment based on how patient is behaving</div> <div>Consider other causes of pain – "Is this your usual sickle pain?"</div> <div>Variability of approach / decision making</div>	<div>Timing of dose</div> <div>Appropriate dose</div> <div>Evaluation of response</div> <div>Route of administration of opioids</div> <div>Role of IV hydratin vs oral</div> <div>Co-administration of tylenol, NSAIs</div> <div>Patient centred care</div> <div>Physician nurse communication</div> <div>Expertise with central venous access devices (ports, PICCs, etc)</div> <div>Experience with baseline long-acting narcotics</div> <div>Prevent constipation</div>	<div>Timing of step down from IV to oral</div> <div>Criteria for step down</div> <div>Additional testing</div> <div>Mobilization</div> <div>Incentive spirometry</div> <div>Psychosocial history / monitoring</div> <div>ED social worker</div>	<div>Involving patients SCD provider(s)</div> <div>Early initiation of d/c planning</div> <div>Role for specialist consultation</div>	<div>Pain plan for discharge</div> <div>Role of SCD clinic vs. family physician</div> <div>Communication with clinic</div> <div>Case management</div> <div>Protected time</div> <div>Psychosocial assessment</div>	<div>Local physician champions /leads</div> <div>ED culture / communication</div> <div>Social work support</div> <div>Team-based settings</div> <div>Links with family practice / primary care / community hospitals</div>
System	<div>Community vs academic setting-expertise, accessibility, patient volumes, history of patient on file—directions for EMSAccess to services</div> <div>Communication with clinical providers eg Rx</div> <div>Patient experience in acute care setting</div> <div>Phone resources: telehealth, SCD clinic / nurse, on call physician</div> <div>Self care plans-up to date, accessible</div> <div>Access to opioid prescriptions (eg no phone prescriptions)</div> <div>Family physician</div> <div>Walk in clinic</div> <div>Day treatment area</div> <div>Hours of operation of outpatient services</div>	<div>Care plan</div> <div>Patient passport / SCD cards</div> <div>EMTs- ODB, clinical records</div> <div>Standardized care pathways / algorithm</div> <div>Triage guidelines (CTAS)</div> <div>Inconsistent triage level-centre dependent (eg patient volume)</div> <div>Lack of access to narcotics at triage</div> <div>Delays in triage due to patient volumes</div>	<div>Evaluation / benchmarks for appropriate time of first dose</div> <div>Patient specific pre-printed orders</div> <div>Short stay unit</div> <div>SCD pain order sets</div> <div>Create a calm environment</div> <div>Culture of care</div> <div>Availability of PCA pumps</div>	<div>Reliable assessment of pain and consistent interval / timing of assessment and evaluation - standardize</div>	<div>Fast track to admission / short stay unit</div> <div>Differences between community and tertiary centres</div> <div>Medical criteria for discharge</div>	<div>Connect patient to comprehensive care outpt. f/u</div> <div>Interval of f/u visit individualized</div> <div>f/u</div> <div>Phone calls</div> <div>Written f/u plan on discharge form</div> <div>Access to fund for small supply of meds on d/c</div>	<div>Outreach programs</div> <div>Designated centres</div> <div>Networks</div> <div>Support groups and advocacy</div> <div>Free access to SCD meds for patients</div>

Focus of the Clinical Practices

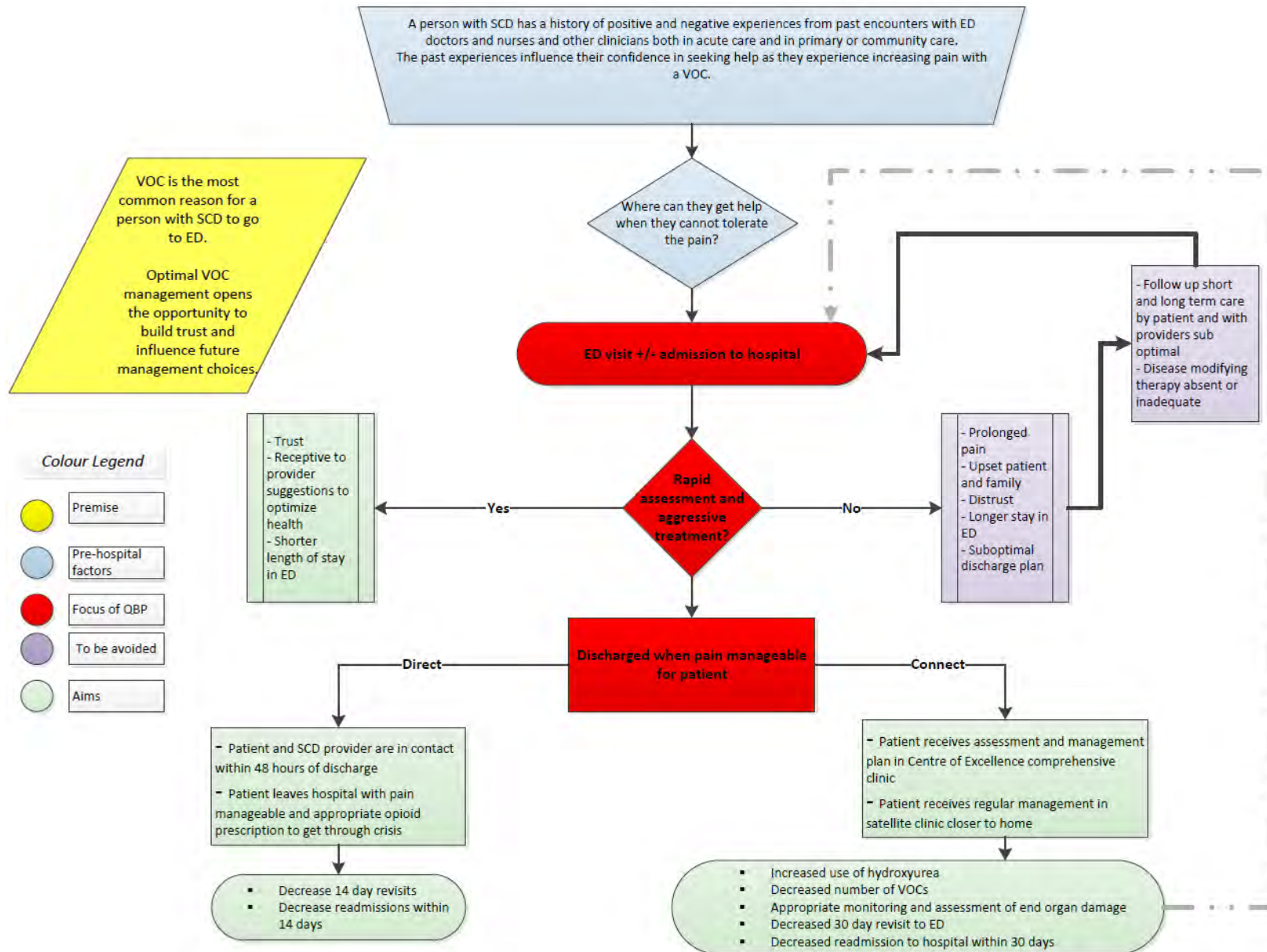
VOCs are the most common reason for emergency department (ED) visits and hospitalizations for patients with SCD. Ensuring that patients receive timely, effective pain relief is one of the most important elements of delivering high quality care to individuals with SCD in the ED.

The **patient's journey** begins with presentation to the ED, with some patients proceeding to admission to hospital, and ends with successful discharge home and a further period of 30 days after leaving hospital (ED or inpatient unit). In other jurisdictions (Florida, Georgia) and in one Ontario pilot project, care that meets best practices has been delivered in environments other than an ED setting. However, the vast majority of initial care in Ontario is delivered in an ED setting. Many hospitals require that any unplanned admission of a patient to hospital occur via the ED. Throughout the best practice recommendations the ED is referred to frequently as that is the current state in Ontario. However it is emphasised that these recommendations are applicable to whatever setting is able to provide the assessment, treatment and monitoring required to treat patients with VOC.

Target Patient Population

The target population are patient with uncomplicated moderate to severe VOC, as VOC without complications of fever/ sepsis, acute chest syndrome, splenic sequestration, stroke or other organ involvement is the most common presentation to the ED. Initial assessment and treatment for VOC occur in the ED, where patients may either be discharged home or admitted to hospital inpatient unit for continuing care.

Figure 3. Recommendations for best practices and their implementation will be described and the following schematic provides the high level view and context for the detailed recommendations.



3.0 Recommending Best Practices¹ in the Management of SCD VOC

Preamble:

1. Team based care in supportive communities and patient/ family education in self-care to prevent and effectively treat VOCs is essential to minimize the use of acute care services.
2. The severity (phenotype) of SCD varies from person to person and some VOCs will not be adequately managed without hospital admission.

Prior to the development of best practices, the Expert Panel members developed a comprehensive overview of the system-, provider- and patient-related issues and gaps across the care pathway of a patient presenting to the ED with a VOC. This mapping process revealed three overarching themes needing addressed to bridge the gaps in the care pathway: education, communication and coordination. (Table 7)

The work of the panel to recommend clinical best practices has been aided by the availability of recent guidelines. The recommendations for best practices for the care of SCD patients with VOC are taken from the following sources:

1. Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014, National Health Lung and Blood Institute
2. Sickle cell acute painful episode: management of an acute painful sickle cell episode in hospital, National Institute for Health and Care Excellence (NICE) Guideline [CG 143], June 2012
3. Consensus Statement on the Care of Patients with Sickle Cell Disease, Canadian Haemoglobinopathy Association (Unpublished) 2014
4. Expert Panel Members
5. Additional evidence based publications as needed

The recommended best practices are presented as:

- a numbered algorithm for care from triage in the ED to discharge (Figure 4)
- a table of recommendations corresponding to the algorithm numbers (Table 8)
 - a focus on the **“Principles of Vaso-occlusive Acute Pain Management”** (Figure 5)
 - an expanded section related to pain management principles and approaches

Excellent algorithms that can be used as clinical tools have been created by others. Such algorithms tend to be brief, use few words and often just address one section of the care path. (Appendix 3) The algorithm created by the SCD VOC QBP Expert Panel was designed to provide a broader view of care and the context within which the care is provided.

¹ Best practice refers to a combination of best available evidence and clinical consensus as recommended by the Clinical Expert Advisory Groups

Figure 4. Algorithm of Care for SCD VOC

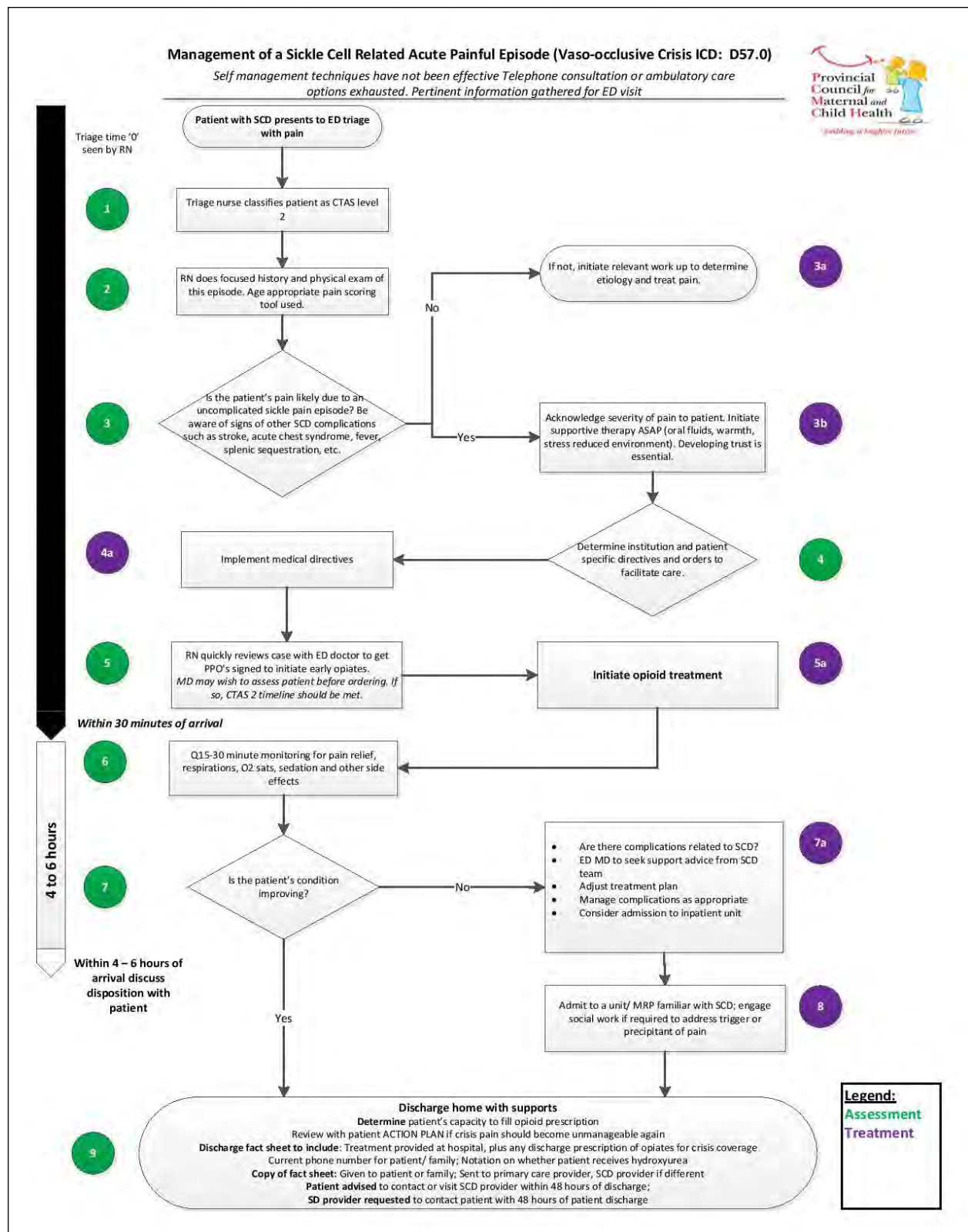


Table 8. Recommendations to correspond with care path algorithm.

	Instruction	Recommendations	Source			Grading
			CCS	NICE	NIHLB	
1	Assign patient CTAS level 2	Treat as an acute medical emergency.		√		Expert Panel consensus
		Patients should be assessed with priority upon arrival, and appropriate management (with opioids) should be started within 30 minutes	√	√	√	<i>Strong Recommendation, High-Quality Evidence (NHLBI)</i>
2	RN Does a focused physical exam and history relating to this episode. Ask the patient:	Is the pain similar to prior vaso-occlusive crises? Does this feel like your usual sickle pain?	√	√	√	
		What are the characteristics, location and intensity of the pain? Any associated symptoms?	√	√	√	Expert Panel consensus
		Assess pain and use an age-appropriate pain scoring tool for all patients presenting at hospital with a VOC.		√		Expert Panel consensus
		Patients with SCD VOC should be cared for in an age-appropriate setting.		√		Expert Panel consensus
		Identify when the pain started and what the patient did to manage the pain? Did they take medications, and if so, record medications, dosage and timing?				Expert Panel consensus
		Does the patient take opiates for chronic pain management related to SCD?		√		QBP Panel consensus
		Clinically assess all patients presenting at Hospital with a VOC including monitoring of: - blood pressure - oxygen saturation on room air by pulse oximetry (if oxygen saturation is 95% or below, offer oxygen therapy to maintain oxygen saturation above 95%) - pulse rate - respiratory rate - temperature.		√		Panel consensus

	Instruction	Recommendations	Source			Grading
			CCS	NICE	NIHLB	
3	Be aware of other SCD complications. Is the pain likely a VOC?	For pregnant women with a VOC, seek advice from the obstetrics team and refer when indicated. Any women 20 weeks gestation or greater should be referred to a high risk obstetrical centre.		√		Expert Panel consensus
		For patients with abdominal pain, examination of the spleen (through palpation) is absolutely necessary to ensure that the pain is not due to acute splenic sequestration.	√			
		Parents of children with SCD must be taught how to palpate for enlargement of the spleen.	√			Expert Panel consensus
		Be aware of other possible complications seen with a VOC, at any time from presentation to discharge, including: - acute stroke - aplastic crisis - priapism - myocardial ischemia	√			Expert Panel consensus
		Patients should be evaluated for constipation upon admission, and appropriate preventative measures should be provided.	√			
3a	Assess for causes of pain unrelated to SCD	Those with atypical pain or symptoms should be evaluated for an alternative etiology for the pain that may not be directly caused by SCD	√	√		Expert Panel consensus
3b	Acknowledge pain and implement supportive measures	Acknowledge the pain Place patient in quietest space possible, away from drafts (and don't forget to check on them) Provide warm blanket or heat pad Developing trust is essential; engage social work if required				Expert Panel consensus

	Instruction	Recommendations	Source			Grading
			CCS	NICE	NIHLB	
		Throughout a VOC, regard the patient (and/or their care-giver) as an expert in their condition, listen to their views and discuss with them: <ul style="list-style-type: none"> - planned treatment regimen for the episode - treatment received during previous episodes - any concerns they may have about the current episode - any psychological and/or social support they may need. 	√			Expert Panel consensus
		Encourage the patient to use their own coping mechanisms (for example, relaxation techniques) for dealing with acute pain.		√		Expert Panel consensus
4, 4a	Determine institution specific and patient specific directives and orders to facilitate care	General medical directives should be in place in the ED to facilitate rapid assessment and preparation for treatment. Additional medical directives specific to VOC events are recommended. Order sets developed collaboratively by clinician experts in SCD, pain management and emergency are recommended to guide best practices. Triage files specific to the patient are recommended for people who attend ED frequently, or in situations where the volume of SCD patients is very low and care is dependent upon the shared knowledge of the SCD provider with the treating physicians. (example: hospitals that have less than 20 ED visits per year for SCD VOC)				Expert Panel consensus
4a	Patients should be started on hydration	Patients should be started on hydration (preferably oral, if tolerated) to ensure that they are not dehydrated.	√			Expert Panel consensus

	Instruction	Recommendations	Source			Grading
			CCS	NICE	NIHLB	
		In euvoletic adults and children with SCD and a VOC who are unable to drink fluids, provide intravenous hydration at no more than maintenance rate to avoid over-hydration.			√	
4, 5	Order sets and medical directives, patient specific care plans and triage files	Once appropriate pain management has been initiated, and depending upon the presenting symptoms, laboratory and/or diagnostic imaging investigations may be considered. Septic work up if fever or symptoms of infection present. No role for chest x-rays or other imaging studies for an uncomplicated VOC. Risk of frequent radiation exposure should be kept in mind. Routine measurement of oxygen saturation by pulse oximetry (SpO2). If further assessment of hypoxemia is indicated (e.g. SpO2 <95%), ABG or capillary blood gases may be performed.				Expert Panel consensus
		Patients should be treated with the most effective therapy with the least potential side effects in each individual patient. The response to treatment should be recorded in each patient's chart so it can be reviewed upon future admissions.	√			Expert Panel consensus
5a		Rapidly initiate treatment with opioids in a VOC with severe pain.	√	√	√	
		The initial treatment should be chosen to achieve appropriate pain control as soon as possible. Patients who have moderate to severe pain or those who were treated with oral morphine without successful pain relief at home should be given intravenous morphine after evaluation.	√			Expert Panel consensus
		Long-acting oral opiates should be considered for basal pain control once an appropriate dose can be approximated if	√			Expert Panel consensus

	Instruction	Recommendations	Source			Grading
			CCS	NICE	NIHLB	
		patients do not have a continuous IV opioid infusion.				
		For breakthrough pain, short-acting medications are preferable over long-acting alternatives. An equi-analgesia chart should be available and used to determine appropriate dosing. Long-acting opioids are generally used for pain that is chronic in nature.	√			Expert Panel consensus
		Patients with adverse reactions to any analgesics should be provided with a written document indicating the type of adverse reaction, so that, in the future, an alternative treatment can be provided as soon as required.	√			Expert Panel consensus
		Hydroxyurea should not be discontinued during a vaso occlusive event unless laboratory results (as per hydroxyurea protocol –) or other indications are present, such as active infection , neutrophils $< 1.0 \times 10^9/L$, reticulocytes $< 80 \times 10^9/L$, platelets $< 80 \times 10^9/L$				Expert Panel consensus
		The response to treatment should be regularly evaluated, and analgesia dose should be reassessed on a regular schedule to titrate the medication dosing to optimize pain control. Step-down therapy should not be attempted until the pain is well controlled.	√	√	√	Expert Panel consensus
		The respiratory status of all patients on infusion of analgesics should be monitored. All patients, especially those with chest or back pain and those with supplemental oxygen requirement, should be started on age-appropriate incentive spirometer.	√			Expert Panel consensus
7a	Reassess for complications, seek support form and SCD specialist, adjust	If the patient does not respond to standard treatment for a VOC, reassess them for the possibility of an alternative diagnosis.		√		Expert Panel consensus

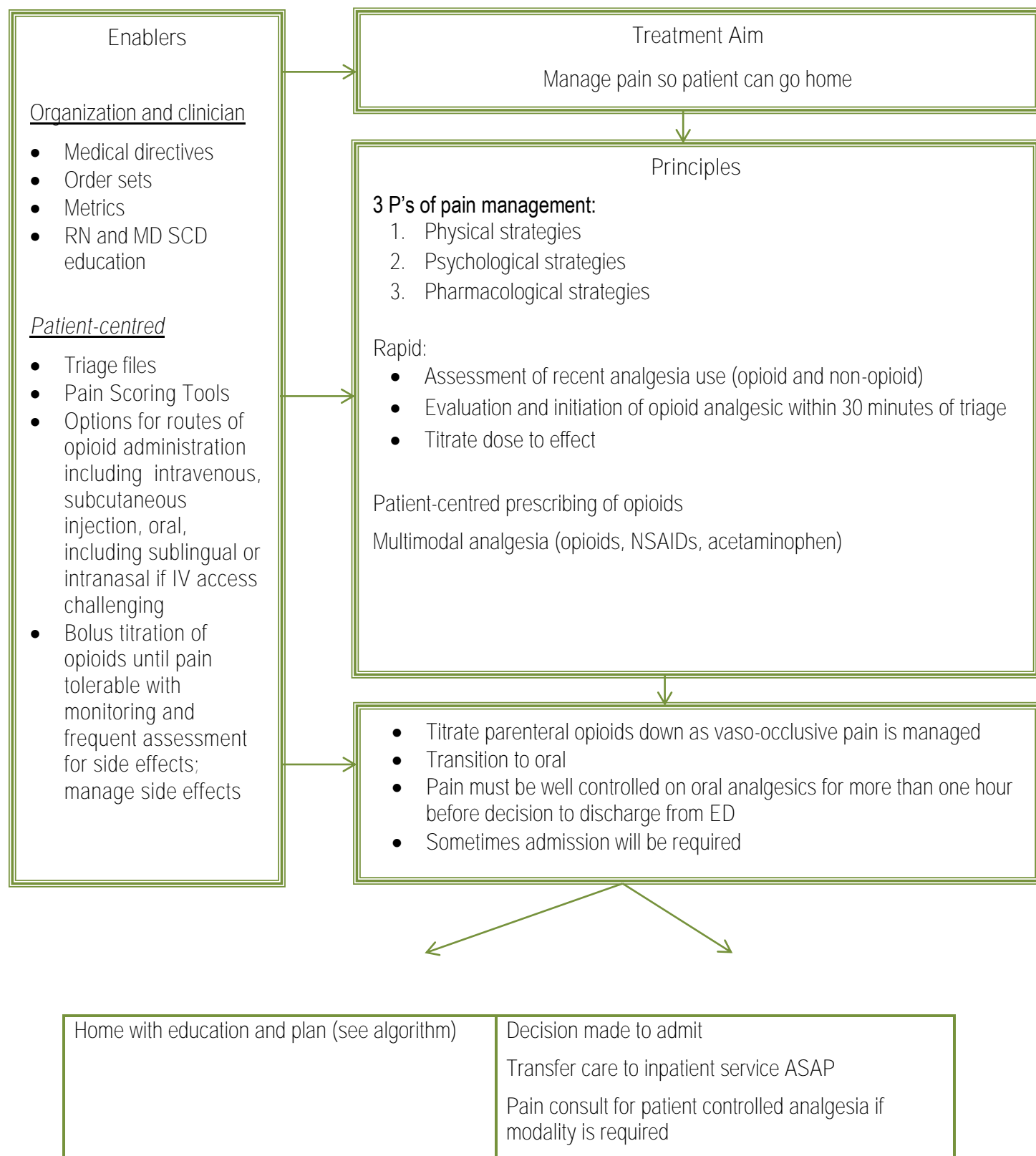
	Instruction	Recommendations	Source			Grading
			CCS	NICE	NHLBI	
	plan, manage complications	For those patients whose pain is not controlled upon infusion of analgesics, a team that is expert in the management of pain (e.g., anesthesia, acute pain service) should be consulted, and PCA or nurse controlled analgesia (NCA) should be started. Once sufficient pain control is achieved, de-escalation of therapy can be attempted.	√			(Moderate Recommendation , Low-Quality Evidence) NHLBI plus Expert Panel consensus
		Optimal treatment of VOC requires a combination of pharmacological, psychological and physical therapeutic approaches and may require expertise from a SCD pain specialist.	√			Expert Panel consensus
		Be aware of other possible complications seen with a VOC, at any time from presentation to discharge, including: <ul style="list-style-type: none"> - acute stroke - aplastic crisis - nfections - osteomyelitis - splenic sequestration (see number 3). 	√			Expert Panel consensus
7		Within 4 to 6 hours of arrival discuss discharge disposition with patient. Patients are best able to identify when they are ready to go home and the pain is such that they can manage it.				Expert Panel consensus
8	Maintain best practices on general medical inpatient unit	<ul style="list-style-type: none"> ▪ Admit to inpatient unit most familiar with SCD VOC management ▪ SCD provider advised of admission ▪ Inpatient most responsible physician to contact SCD expert (if patient's SCD provider not available to discuss ED care) Incentive spirometry recommended during inpatient stay to prevent evolution of acute chest syndrome.				Expert Panel consensus

	Instruction	Recommendations	Source			Grading
			CCS	NICE	NIHLB	
		Pain medication to be ordered at regular intervals (not only as required) with appropriate drug/dosing.				
		Provide non-pharmacological measures such as massage, heat pad, and distraction. Involve a pediatric life specialist for children. Encourage activities as tolerated.	√			Expert Panel consensus
9		Discharge summary to include: <ul style="list-style-type: none"> • Treatment provided at hospital, plus any discharge prescription of opiates for VOC coverage • Current phone number for patient / family Notation on whether patient receives hydroxyurea Copy of summary: <ul style="list-style-type: none"> • Given to patient or family • Sent to primary care provider, SCD provider if different Determine patient's capacity to fill opioid prescription				Expert Panel consensus
		Review with patient what to do if pain should become unmanageable again <ul style="list-style-type: none"> • Patient advised to contact or visit SCD provider within 48 hours of discharge; SCD provider requested to contact patient within 48 hours of patient discharge 				Expert Panel consensus
Not recommended						
	Do not offer pethidine (demerol) for treating pain in a VOC			√		Expert Panel consensus
	Inhaled nitric oxide, corticosteroids, or magnesium sulfate have not been shown to be effective during VOC.		√			Expert Panel consensus

	Instruction	Recommendations	Source			Grading
			CCS	NICE	NIHLB	
		Transfusion of episodic packed red blood cells for the management of uncomplicated VOC not recommended; however, it can be given for the management of symptomatic anemia. Red cell transfusions do not have a beneficial role in the management of VOC in SCD. The benefit of chronic transfusion therapy for the management of chronic sickle cell pain is controversial	✓	✓		Expert Panel consensus
		Transfuse only with the approval of haematologist				Expert Panel consensus
		Antibiotic therapy has no role in the management of a simple VOC, and when there is no fever or any other findings suggestive of infection. Physicians should be aware that patients with SCD may have a high baseline WBC count, and leukocytosis in isolation may not be due to infection. Nucleated red blood cells can also be erroneously counted as WBCs on automated CBCs – manual WBC count and WBC differential are often required.	✓			Expert Panel consensus
		No routine intravenous fluid bolus in patients with VOC unless clinically indicated.				Expert Panel consensus

Expanding on Algorithm point 5a: Initiate Opioids

Figure 5. Principles of Vaso-occlusive Crisis Management



Pain Management Best Practices

Analgesic history:

- ✓ The patient should be well prepared with education in self-management and should have a care plan for managing early stage VOC.
- ✓ The patient should bring their medications and record of self-administration with them to ED.

3 P's of Pain Management:

- ✓ Physical pain techniques (e.g., massage, warm blankets / heat packs to the affected area, stretcher adjusted to position of comfort, quiet surroundings, etc.).
- ✓ Psychological techniques (e.g., emotional support, behavior management, distraction with music, video games, TV, etc); minimization of psychological stress related to provider interactions/environment (e.g., behaviours that are not empathetic add to psychological stress which makes it more difficult to manage pain).
- ✓ Pharmacological treatment and hydration are the cornerstones of the management of VOC.

Principles of Acute Pain Management

- ✓ Rapid assessment to establish that the patient is having a VOC .
- ✓ Aggressive treatment with opioids at the right dose, the latter to be determined by: (a) whether the patient has taken opioids at home to manage this episode, (b) whether the patient has chronic pain managed by opioids, and (c) the dosages that the patient has had in the ED in the past to manage their pain.
- ✓ Titrate opioids until effective pain relief has been achieved, as measured by a validated pain assessment tool. Intravenous bolus delivery is optimal to initiate treatment and achieve rapid pain relief, with repeat doses every 15-20 minutes (while assessing for pain relief and sedation). Can initiate opioid therapy using any other route (eg. Oral, SC, intranasal) if immediate IV access not available)
- ✓ Patient-centred pain management is reflected by **rapid, aggressive use of the 'opioid of choice'** for the patient. Base analgesia selection on pain assessment, associated symptoms, outpatient analgesic use, patient knowledge of effective agents and doses, and past experience with side effects. There is NO standard drug, dose, route or frequency of administration that is applicable to/effective for ALL patients.
- ✓ **Multimodal pain management.** *"This approach simultaneously administers two or more analgesic agents with different mechanisms of action. Combination therapy using drugs with distinct mechanisms of action may add analgesia or have a synergistic effect and allow for better analgesia with the use of lower doses of a given medication than if the drug were used alone^{iv}."*(Pasero 2011)

I know what I need. I've been going through this for years. It may not be the standard dose, but I need it."

Analgesics and adjuvants

The effectiveness of analgesics varies from person to person. Treatment should include a standard opioid approach with options available for patients who have had reactions to one opioid or have had better results from one opioid over another. Non-steroidal anti-inflammatory medications (NSAIDs) such as ibuprofen at home and ketorolac (parenterally) in hospital are recommended as adjuvant medications in the absence of contraindications. Acetaminophen can be added as well if there are no contraindications. Over time people with SCD develop chronic disease related to tissue and organ damage; some medications will not be recommended when specific organ damage is present. Manage side effects, most commonly nausea, pruritus and sedation. Be aware of further sedative effect from medication used to manage side effects e.g. dimenhydrinate, diphenhydramine.

Dose

Titrated intravenous bolusing of opioids to effect is recommended. Delivering frequent (every 15- 20 minutes) boluses in the initial phases of treatment in acute care (usually the ED) provides a rapid systemic effect. The patient should be monitored as per organization policy following the administration of opioids. The goal for patient care is that they receive prompt attention, safe and effective pain management. Once pain control has been achieved, opioids can then be given in regularly scheduled doses aimed to provide longer-term analgesia (e.g. oral long-acting formulations, transdermal patches or continuous IV infusions) with appropriate orders for breakthrough dosing. The evidence supports this aggressive management of acute pain^v. Attention must be given to equivalence when converting route or drug. Emergency department length of stay among those with SCD VOC has been shown to be reduced with rapid, aggressive pain management with opioids. Dose ranges should be standardized for the opiate naïve. People with SCD can develop chronic pain and their requirements might be much higher than with opioid naïve patients.

"I have a routine- hot shower, fluids, rest, my family will massage me, but after 24 hours if the pain moves to another place I know I have to get to the hospital. I've had this all my life, I have no veins left and I tell them that and they say 'oh we have someone who is really good (at starting IVs)' and an hour later I've had 12 pokes and no pain medication, not even anything orally."

Route

Options for route of administration should be identified for analgesics. Intravenous delivery of opioids is optimal for the initial management of acute pain in the ED; however at times difficulties starting an intravenous can occur. This should not delay opioid administration. Alternate delivery can provide bridge dosing while a clinician with the expertise to place the intravenous catheter is located and arrives at the bedside. Intranasal, subcutaneous and oral, including sublingual routes should all be considered.

Patients may present with central venous access catheters in place; ED clinicians must have access to clinicians with expertise in working with these lines. While waiting for support, alternate routes of opioid administration should be used to provide bridge dosing.

Patient controlled analgesia (PCA) is recommended for patients who are competent to manage PCA and are in environments where clinicians are competent and able to initiate PCA. Oral PCA has been used in the appropriate environments with patients who are carefully selected for their ability to take their analgesic as required^{vi} and to record their utilization. Both of these patient controlled options require careful patient selection, clinician education, resources and processes in place to ensure safety.

Organizations that have long waits from discharge disposition to transfer to an inpatient bed should ensure transfer of care to an inpatient physician / service as quickly as possible. In the interim, the emergency physician should write standing orders for analgesia during waiting time of transfer of care to in-patient consultant to avoid lengthy times of no analgesia. A process should be developed to support patients with the delivery of pain medication and monitoring in the least noisy and stressful environment possible, reducing negative **environmental impact on the patient's ability to rest and sleep** while awaiting consultation and transfer to an in-patient bed.

*"They put you on a stretcher and leave you there. The nurses glance in and walk by. They don't want to make eye contact with you."
"I had to call my family to say no one was helping me to go to the washroom. You go in with the simplest things and you can end up with far worse. It's like, If I continue to go to this hospital, they're going to kill me."
"It can be isolating and it can be degrading. There are times I can't go to the bathroom by myself and really need someone to help me."*

Many clinical pathway algorithms, order sets and medical directives (both adult and paediatric) have been developed for the management of SCD patients with a VOC. Organizations are encouraged to liaise with their nearest Centre of Excellence or Satellite Centre to obtain examples.

Patient Self-Management

People with SCD, and their families, need education on how to recognize the onset of a VOC as well as the physical, psychological and pharmacological self-**management measures to take**. **Understanding one's** own pattern and experiences in order to manage them is a critical component of living with SCD. Patients are encouraged to keep a pain diary and have an action plan should the pain become unmanageable at home. Patients should be educated about common triggers for VOC, and work with members of the healthcare team to develop prevention strategies.

All patients with SCD should have a home care / action plan developed in conjunction with their SCD provider. The plan should include:

- A reminder of warning symptoms that should prompt urgent ED/SCD team care (fever, chest pain, shortness of breath, signs of stroke, splenic enlargement, etc.)
- Steps to manage pain, analgesia type, dose and frequency, hydration, warm blankets or warming pads, quiet and a stress free environment
- Support of family member or advocate
- Contact information for a SCD provider for advice and consultation
- Plan on how to travel to their regular hospital
- Details of the VOC, including what treatments have been implemented (i.e. the name, dose and timing of oral pain medications at home), should be documented and brought to hospital if it becomes necessary.

Providers should understand that VOC can and will occur despite informed and conscientious self-management.

"Doctors and nurses need to be more educated. As well on our side we need to educate ourselves, what can help, we need to take care of ourselves."

4.0 Implementation of Best Practices

The relatively small population of Ontarians who have SCD live for the most part in the Greater Toronto Area (GTA), Ottawa and Hamilton. The rates of ED visits, repeat ED visits, hospital admissions with return to ED or readmissions after discharge are high. Literature demonstrates that prompt and aggressive treatment decreases the length of stay in the ED. Fourteen day ED revisits or readmissions to hospital are associated with care received during ED visit or admission while 30 day revisits or readmits are associated with care received in the community.

Achieving best practices for the management of acute pain of a VOC, for all patients, no matter where they live will require a system level strategy with multiple approaches to strengthen the performance of all actors within the system. These approaches must tap into the existing health resource capacity and address the system-, provider- and patient-related gaps at all points of care delivery with measurable indicators by which performance will be monitored.

Literature on process and models that impact frequency of ED visits and hospital admissions

Paediatric 30 day readmission to hospital related to SCD VOC is related most strongly to lack of outpatient follow up with haematologist, as well as a diagnosis of asthma or having been on room air for less than or equal to 24 hours after having required supplemental oxygen to maintain oxygen status^{vii}.

Studies^{viii, ix} also show that readmission rates for patients with SCD further rise with age, peaking in the 18-30 year age range with rates of 40%. This very age group, when transition to adult care usually occurs in North America, is associated with increased mortality within this patient population^x.

In adults, a before and after evaluation of the provision of care within a clinic employing a chronic care model² showed a significant increase in the use of hydroxyurea, a trend toward decreased hospital admissions per patient per year and a decrease in 30 day readmissions. This evaluated intervention took place in a tertiary setting with comprehensive educational efforts across ED and inpatient units by all team members. The care team included a medical internist expert in the care of people with SCD, a nurse practitioner, a social worker and patients had 24/7 telephone access to a knowledgeable care provider for coaching through the early stages of a VOC^{xi}.

The difference between paediatrics and adults and the subsequent research focus is twofold:

- **the advocacy of parents to provide much of the 'coordination' in children's care**
- in adult years the chronicity and complexity coupled with the lack of parental advocacy require that care be optimized through the chronic care model

The evidence is endorsed by the clinical experts on the need to see a SCD provider soon after hospital encounter for VOC and the use of the chronic/comprehensive care model is required as chronicity sets in.

Evidence continues to indicate that quality improvements that further reduce ED revisits, frequency of VOC, admissions and readmissions include the implementation of individualized, multimodal treatment plans that

²² The chronic care model, as described by Wagner, has 6 components including: (1) community resources and policies, (2) health care organization, (3) self-management support, (4) delivery system design, (5) decision support, and (6) clinical information systems.

cross ED, inpatient and outpatient care settings^{xii}. Even for the higher users in the 18 to 30 year old age group, which a large population based study across eight American states identified as having a readmission rate of 41.1 percent within 30 days and 28.4 percent within 14 days^{xiii}, reductions in acute care utilization can be achieved.

System Level Tactics to Enable Best Practices

Implementation recommendation number 2

Formalize ‘Centres of Excellence’ with dedicated funding, at the following paediatric and adult

Academic Health Science Centres:

- a. McMaster / Hamilton Health Sciences Centre
- b. SickKids
- c. University Health Network / Toronto General
- d. The Ottawa Hospital
- e. **Children’s Hospital of Eastern Ontario**

Implementation recommendation number 3

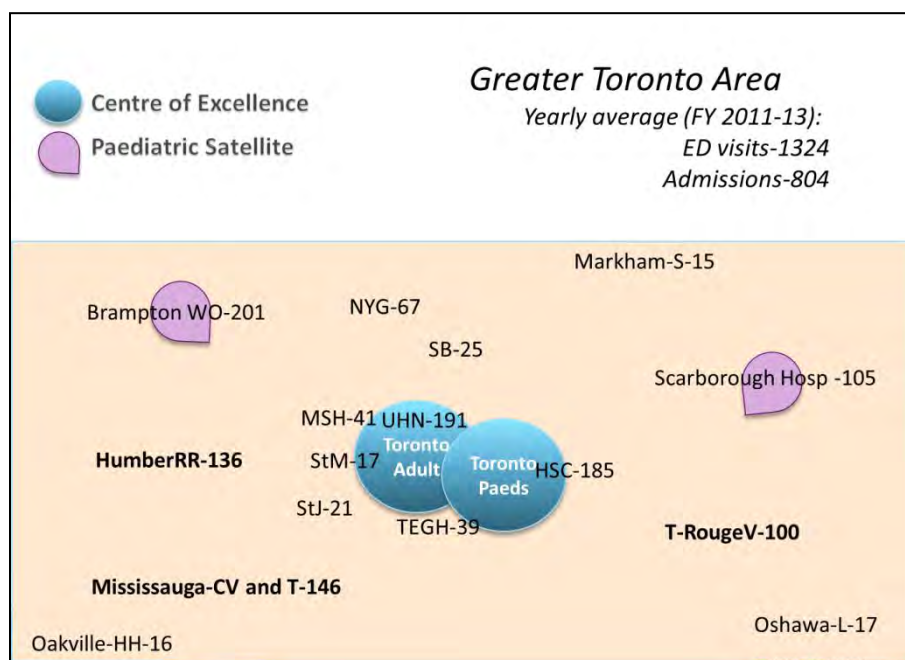
Develop adult and paediatric satellite centres at high volume hospitals within the GTA, modelled on the existing paediatric satellite Centres at William Osler and Scarborough Hospitals. The newborn screening data indicate that these volumes will continue to grow.

Implementation recommendation number 4

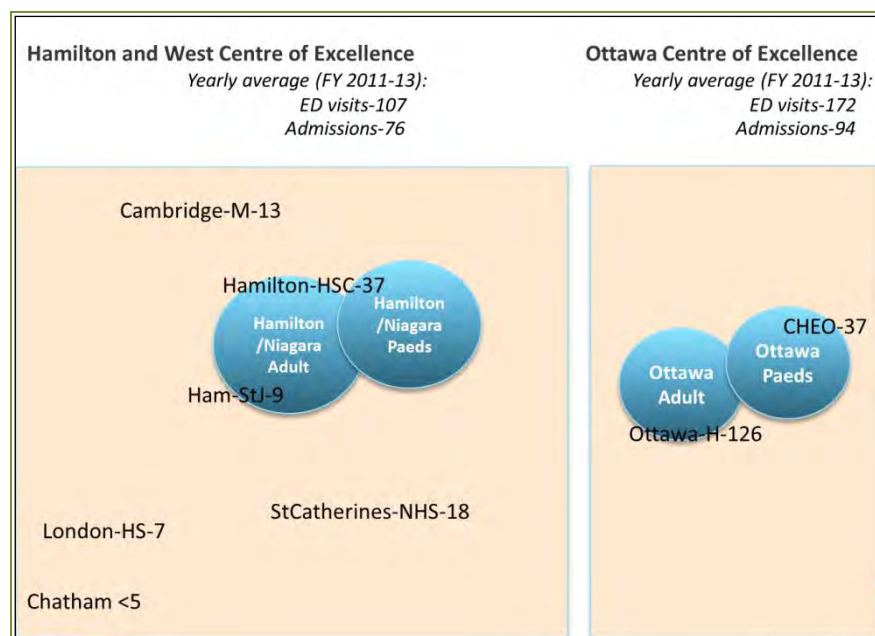
Formalize a hub and spoke relationship between the Centres of Excellence and Satellite Centres.

A schematic (Figure 3) has been developed to illustrate of GTA hospitals, existing Centres of Excellence, existing and recommended Satellites and other high volume sites (Humber River and Mississauga) where both paediatric and adult satellite centres should be developed.

Figure 6. Hub and Spoke of Centres of Excellence and Satellite Centre



The numbers are the yearly average of ED visits at hospitals in LHINs 5 to 9 (FY 2011-2013)



The numbers are the yearly average of ED visits at hospitals in LHINs 1-4 and LHINs 10 and 11 (FY 2011-2013)

A hub and spoke model provides the structure to support knowledge translation and care delivery closer to home. In order for the hub and spoke model to successfully create the conditions to drive and sustain best practices the roles (structures and processes) that each organization fulfills must be clearly articulated.

Centres of Excellence

- All people with SCD linked to a C of E.
- Participates in and facilitates basic and clinical research
- Participates in research and evaluation to improve quality of care
- Liaises with all providers involved in the care of people with SCD within the region served
- Develops and delivers education aimed at providers who care for people with SCD, with particular emphasis on Satellite teams as partners
- Provides on call SCD expertise to support physicians caring for people with SCD within the C of E or at another acute care facility within the region; promotes service to low to moderate volume hospitals
- Develops and shares educational materials for patients and families^{xiv}

Roles

- Haematologist
- Clinician focused on pain management (could be Nurse Practitioner supported by pain team)
- Social worker
- Neuropsychologist
- Nurse Practitioner / Nurses

Resources

- Emergency Departments
- Pain services
- Critical care services
- Located in or has access to diagnostic capabilities to monitor organ damage related to SCD

Centre of Excellence Approach

- Models organization wide standardization in care
- Developed order sets in collaboration with ED, haematology, inpatient service, pain services (including pharmacy)
- Identifies validated age appropriate pain measurement tools and ensures they are integrated into practice in all care settings
- Formalizes transition from paediatrics to adult health services preparation^{xv}
- Individualized case management approach for people who are high users of acute services; partner with team-based community providers to enhance community supports (CHCs, CCACs, HealthLinks)
- Clinical expertise 24/7 provided in-house and available to community specialists
- Automated red cell exchange
- MRI
- Other specialists with understanding of SCD

Regional Paediatric and Adult Centres of Excellence Relationships with Each Other:

- Collaborate on regional quality initiatives and outreach activities
- Collaborate to develop regional strategies to resolve issues affecting both adult and paediatric populations (e.g. central access, uptake of hydroxyurea treatment)
- Includes satellite teams in development and delivery of clinician CMEs (rounds, conference sessions, seminars)

Implementation recommendation number 5

Implement a formal transition process using evidence-based readiness and (when available) risk assessment tools^{xvi}

- Collectively the C of **E's and Satellites in Ontario** formalize a Network Relationship/Community of Practice to increase the quality of care for people with SCD across Ontario; specifically develop communication materials to be provided to patients at any point of contact in the health care system regarding:
 - hydroxyurea therapy
 - self-care measures to be taken at the earliest awareness of onset of a VOC.

Satellite Centres

- Serve high volume communities
- Link to C of E for shared care, consultation, education
- Designated Satellites to offer both paediatric and adult programs
- Satellites formally appointed program leads (both clinical and administrative)
- SCD teams developed and lead practice within the organization
- Teams to include ED MDs and RNs, paediatricians, haematologists, anaesthetists/pain clinician, social worker, case manager for intensive support where needed (some team members consult as needed)
- Participate in CME/all clinician education locally and with C of E (consider OTN access)
- Maintain patient referral and physician consult relationship with C of E and encourage all people with SCD to be seen in a C of E at least annually.
- Cross-setting (ED, inpatient) protocols, medical directives, order sets, access to SCD expertise
- Monitor metrics and indicators; QI and process improvement to meet indicator targets

System level approaches to link the many organizations that provide care is essential to ensure sustainable quality care for VOC and SCD more broadly. Relationship building between providers, programs and organizations is the first step and must be formalized in a network of SCD provider-leaders. Shared educational resources and collaboration to develop educational resources will provide some economy of scale and allow Satellite Centres to benefit from the capacities of Centres of Excellence. Most community hospitals in the GTA have ED visit and admission volumes to suggest that the organization should develop a Satellite Centre or at the very least formalize a programmatic approach and leadership to implement best practices.

e-Health

System level electronic solutions for and communication of patient disease state and medication history across organizations have been tried but have not made a significant and sustainable difference in quality of care.

The ease and speed of use and resulting acceptance by clinicians limit the degree to which current electronic solutions are significant levers in quality improvement immediate to the point of care. They do provide data capture monitoring of quality and improvement. In the future the regional Electronic Health Records may be a useful tool.

Organization level

Implementation recommendation number 6

Assign leadership and resources to the Centre of Excellence, Satellite Centre or Sickle Cell Program as appropriate to the role your organization should take within the hub and spoke system of sickle cell care.

Implementation recommendation number 7

Determine the quality improvement approach that the organization will take to enable best practices to be delivered.

Inter-disciplinary collaboration among the ED clinicians, haematologists (adult and paediatric) and pain specialists is essential to developing solutions that incorporate the expertise of each discipline in the clinical solutions (evidence based and organizationally practical).

Implementation recommendation number 8

Approaches to deliver best practice must coordinate care across ED and the inpatient unit.

The clinical processes and clinical flow pathways that must be put in place to achieve the rapid and effective treatment of VOC must be identified within each organization. The barriers to care (knowledge, physical space, policy, interdisciplinary silos) must be identified and solutions developed.

Enabling VOC Management Best Practices³

Implementation recommendation number 9

Expedite care with medical directives that enable nurses to fast-track assessment and prepare for pain management.

Implementation recommendation number 10

Implement best practices beginning with pre-printed order sets that guide clinicians to order and carry out evidence-based care. Hospitals that care for both adults and children with SCD should have separate order sets for adult and paediatric care. Interdisciplinary development of order sets is highly recommended.

³ These enabling recommendations are consistent directions within Accreditation Canada's Required Organizational Practices Handbook 2016 (<http://www.accreditation.ca/sites/default/files/rop-handbook-2016-en.pdf>) and the Institute for Safe Medication Practices guidance for high-alert medications (<http://www.ismp.org/Newsletters/acutecare/showarticle.aspx?id=45>)

Implementation recommendation number 11

Pain assessments should always be made with age appropriate validated pain tools. (Appendix4)

Patient-centred care will require that when patients need specific opioids that are not the opioids most commonly used, these medications are available and administered in the right dose and through the **best available route**. All EDs should have an 'Opioid Equianalgesic Chart' available, supported and endorsed by the organization's pharmacy and pain team^{xvii}.

Implementation recommendation number 12

Patient-specific order sets and triage files are recommended, especially for the frequent users of ED care, people with underlying chronic pain or any other condition that makes their VOC pain treatment vary from the norm. Also consider using patient-specific order sets in hospitals with very low volume of SCD patients.

Implementation recommendation number 13

Set targets and monitor quality improvement metrics internally. In addition to provincially available quality indicators developed using data submitted to Canadian Institute for Health Information, organizations wanting to monitor and improve the quality of acute pain management for patients experiencing a SCD VOC are advised to focus on the three organization level metrics:

- a. The length of time from triage to administration of the first dose of opioids.
- b. The time to an initial reduction of pain score that is meaningful to the patient, e.g. 2 points on a 0 to 10 scale
- c. The length of time from triage to a pain score reduction that is sufficient for the patient to be able to manage the pain at home.
- d. Patient experience data, specifically if available, the three questions in the Picker instrument related to pain management.

For all aspects of care from assessment to treatment and monitoring, the importance of medical directives to expedite care and preprinted order sets to guide best practices have been strongly recommended. These are not simple 'make them and forget them' solutions. The medical directive developed by CHEO for the ED nurses to expedite **care**, "Investigation/initiation of intravenous access on immune-compromised patients with fever by nurses in the **Emergency Department**" documents the following quality management process within the directive:

- All RNs will be educated by the clinical educator or delegate about the purpose, indications and contraindications of initiating the investigations and interventions outlined in this medical directive.
- All incident reports related to initiating investigations under this medical directive will be reviewed by the Clinical Manager, Medical Director and physician involved with care of the patient.
- Audits will be conducted periodically to ensure that the medical directive is being used appropriately.

Likewise, preprinted order sets need to be developed and owned by the end users and medical and clinical supervisors, and be part of a multi-modal intervention to improve care. A publication (Frei-Jones, 2009) on a multi modal intervention for patients admitted with SCD VOC showed reduced rates of readmission within 30 days. The interventions included a nurse who educated family and caregivers, preprinted orders and a third intervention, the addition of regular physician education on SCD and how to use the orders. When the regular physician education was discontinued the 30-day readmission rates increased.

Quality measures such as use of PCA analgesia, greater change in pain scores by the time of discharge and the booking and keeping of a follow-up haematology appointment were improved with the use of the preprinted orders ^{xviii}.

Patients who attend ED frequently, most often have chronic disease related to damage from SCD and/or chronic pain treated with opioids. They attend ED for VOC and are best served during the VOC with individualized patient-specific orders or triage files. These documents may include a description of the patient's disease state, identify the medications and dosages that have worked for that patient in the past and recommend that their haematologist or the acute pain team and social work or a case manager be **contacted immediately upon the patient's presentation at ED**. Organizations may need to develop referral relationships for chronic pain management to chronic pain management programs.

Research in the physiology of chronic pain of musculoskeletal origin has **demonstrated that 'normal' physiologic responses to pain such as peaks in blood pressure and heart rate may not occur and endogenous opioids may not have the same pain relieving effects**. Adults with SCD often note that the nurses and doctors do not believe they have the level of pain they are reporting, particularly because they may not be showing physical signs of painful distress. **Believe the patient's identification of pain severity.**

Implementation recommendation number 14

Clinicians must be educated that in this population the best assessment of severity of pain is the patient's.

Intensive disease and chronic pain management and social work interventions are required to mitigate the need for ED visits in the high user group. For many patients with SCD, repeated poorly treated VOC results in deteriorating quality and quantity of life.

Medical directive and pre-printed orders should be in place to expedite care and support best practices across the organization and personalized plans are recommended for the high users of acute services.

One pre-hospital enabler is recommended to expedite rapid ED triage and treatment: the paramedics call the hospital where the patient will be taken to notify the ED that a SCD patient with acute pain is en route.

In summary, the clinical protocols, medical directives, preprinted orders, patient-specific orders or triage files are essential enablers for best practice. But, to make the desired impact, these enablers must be combined with clinician education and practice monitoring and evaluation through focused audits. These clinical process improvements should be designed to work within the existing work-flow and process to ensure consistent and sustainable quality care delivery. Process or work-flow redesign, where required, must include all involved, from registration clerk, to ED nurses and doctors and any others in or beyond the ED involved in the care of the patient with a VOC.

"In the 1980's there was a protocol in a huge binder in the ED. If they looked at the protocol they gave me exactly what I needed. When the doctor no longer worked at that hospital, they would not use the protocol."

"Best thing is to stay at one hospital so they have all the notes."

"They do not follow the notes that are on file."

Several approaches including Day Units and other models based on inter-professional collaboration between ED staff, haematologists and pain service staff have been developed and implemented in other jurisdictions (Example- Appendix 5). In Ontario, the University Health Network (UHN) are piloting and evaluating a program in which the patient calls the SCD Program Nurse Practitioner (NP) when they develop a VOC. The NP uses a protocol to assess them to ensure the VOC is not associated with related complications and if not, directs them to the Acute Care Ambulatory Unit (ACAU) who relays it to the nurses to expect the patient. The UHN leadership provided education on the assessment and management for people having a VOC to the staff of the ACAU.

The assessment service is available from Monday to Thursdays from 8:00 – 16:00, but upon arrival at the ACAU the patient can remain overnight if needed. While early metrics on patient satisfaction, treatment outcomes and length of stay are positive, a considerable number of patients prefer to attend an ED in their own community^{xix}.

A simple guide that aligns with the recommendations of this report and follows the principles of quality improvement has been recently released by **The National Institute of Child's Health Quality**, "Sickle Cell Pain in the Emergency Department: A Guide to Improving Care"^{xx}.

Provider level

Implementation recommendation number 15

It should be recognized that patients with SCD presenting with acute pain from VOC should be assigned and treated as CTAS level 2 and be given emergent care for their pain. This is a fundamental right of the patient.

Implementation recommendation number 16

Extensive education on best practices for nurses and doctors who work in hospitals with high volumes of ED visits of people in acute pain from a SCD VOC will be required.

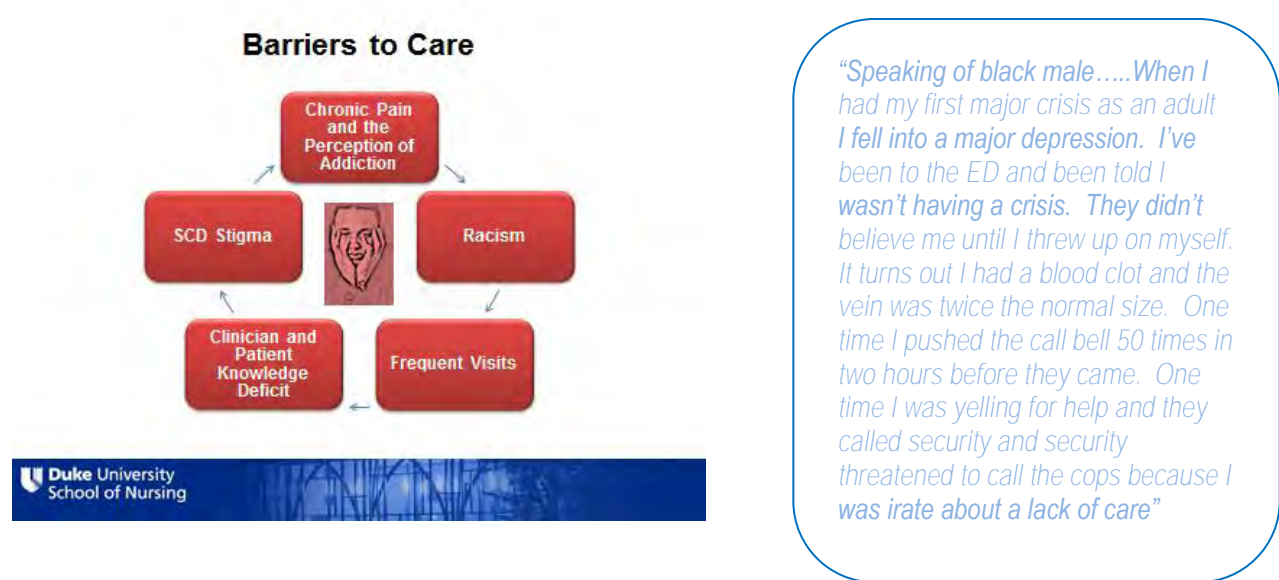
Educate To Overcome Attitudinal Barriers to Care

Education for providers must also address attitudes that are barriers to care. Information must be shared to help providers understand the strength of evidence in support of the unacceptably long wait times for analgesia by people with SCD VOC. Timeliness of initiation of analgesic therapy should be paramount in recommendations to implement best practices within hospital settings.

Clinicians and administrators recognize that racism towards patients with SCD does exist and must be addressed in education for clinicians. Racial bias, whether the clinician is aware of it or not, negatively impacts patient-provider communications.

*"There is a visual disconnect. You are not bleeding or broken."
"ED is for emergencies. It is an emergency...that is why I am here."
"Yes I look this way but you cannot see my pain."*

The potential for a reinforcement cycle between the illness, racism and providers is well depicted in the following graphic:



While clinicians cannot control all of the factors that negatively affect people with SCD, they can recognize and avoid contributing to the 'reinforcement cycle' of negative experiences and responses.

Table 9. Summary of implementation recommendations

Number	Recommendation
1	A communication is developed to clarify for coders and physicians the intended use of the sickle cell crisis code D 57.0.
2	Formalize 'Centres of Excellence' at the recommended paediatric and adult Academic Health Science Centres.
3	Develop adult and paediatric satellite centres at high volume hospitals with in the GTA, modelled on the paediatric satellite Centres at William Osler and Scarborough Hospital.
4	Formalize a hub and spoke relationship between the Centres of Excellence and Satellite Centres.
5	Implement a formal transition process using evidence based readiness and (when available) risk assessment tools.
6	Assign leadership and resources to the Centre of Excellence, Satellite Centre or Sickle Cell Program as appropriate to the role your organization should take within the hub and spoke system of sickle cell care.
7	Determine the quality improvement approach that the organization will take to enable best practices to be delivered.
8	Coordinate approaches to deliver best practice across ED and the inpatient unit.
9	Expedite care with medical directives that enable nurses in fast tracking assessment and preparing for pain management.
10	Implement best practices beginning with preprinted order sets that guide clinicians to order and carry out evidence based care.
11	Pain assessments should always be made with age appropriate validated pain tools.
12	Patient-specific orders sets and triage files are recommended, especially for the frequent users of ED care, people with underlying chronic pain or any other condition that makes their VOC pain treatment vary from the norm.
13	Set targets and monitor quality improvement metrics internally.
14	Clinicians must be educated that in this population the best assessment of severity of pain is the patient's.
15	Recognize the right and validity of people with acute pain from a VOC to be treated as CTAS level 2, emergent care needed.
16	Extensive education on best practices for nurses and doctors who work in hospitals with high volumes of ED visits of people in acute pain from a SCD VOC will be required.
17	Formally create a network of provider leaders, many that are currently in the Expert Panel, as an advisory body within PCMCH (or another secretariat or agency), mandated by the Ministry to oversee and monitor implementation of quality improvement initiatives to enable best practices.

5.0 What Does it Mean for Multi-disciplinary Teams?

ED/Inpatient Care

Nurses and physicians must

- be provided and take educational opportunities to increase their knowledge and skill in delivering care to the patient having a VOC.
- receive education to understand the behaviours that reflect prejudice or any lack of caring to people having a VOC.

Nurses

- must have medical directives to expedite assessment and early treatment measures. Nurse educators must provide the front-line nurses with the education to knowledgeably execute the medical directives.
- through education and the use of clinical supports (tools), demonstrate knowledge and competence in assessing and expediting care so that physicians trust their assessments and can expedite the physician component of timely care.

Physicians

- must through education and through the use of clinical supports (tools) demonstrate knowledge and competence in providing care that adheres to best practices
- adult and paediatric physicians from ED, haematology and pain services must collaboratively develop order sets (different for children and adults).

Nurses, physicians, administrators and ED registration staff must participate in the development of processes that eliminate barriers and are manageable within current work flow processes. Where teams feel this is not possible or desirable to achieve the standards of care in the existing work flow, then new approaches must be developed.

Relationships must be developed between internal disciplines across units in an organization. ED physicians and nurses must collaborate with haematologists and the most responsible physicians on inpatient medical units where the patients are admitted to develop coordinated order sets so that treatment approaches are consistent. If patients receive care in haematology clinics in the organizations, the team from those clinics should also be part of developing and ensuring consistent treatment approaches.

Transition

The paediatric team, the patient and their family and the adult SCD team must develop formal transition preparation processes for handing over paediatric patients to adult receiving teams. This piece of work will be critical in building knowledge capacity among adult providers and raising the standards of care in the adult settings.

6.0 Support for Change

Implementation of recommendation number 17

Recommend a network of provider leaders, many that are currently in the Expert Panel, formally create an advisory body within PCMCH (or another secretariat or agency), mandated by the Ministry to oversee and monitor implementation of quality improvement initiatives to enable best practices. The advisory, **through PCMCH's** (or other) program development and management capacity will provide mentorship and coaching to support sites in quality improvement initiatives to meet indicator targets through best practices. The advisory will interpret performance metrics to support PCMCH in performance monitoring.

- The advisory should develop a comprehensive plan with costing to develop and sustain a hub and spoke system that will include 5 to 7 additional adult Satellite Centres and 2 to 3 additional paediatric Satellite centres.
- The paediatric and adult Centres of Excellence will formalize collaborative efforts in achieving comparable standards of care that are reflective of best practice recommendations and make transitions in care from paediatric to adult health services less distressing.
- Centres of Excellence and Satellite centres shall establish provider- leader relationships and collaboratively plan shared education and share educational materials and resources

7.0 Afterword: Beyond the Walls of Acute Care: Home and Community

Home

When one family member has SCD the whole family is impacted. Unplanned visits to ED, hospitalizations, and chronic pain impact school and the work place and bring chronic and intermittent peaks of stress to families. Strong family, community and health team supports both in primary and specialty care settings are essential to help people with SCD, and their families optimize their health and overall quality of life.

Community Care

In Ontario, the establishment Community Health Centres (CHCs), in both rural and urban settings has been associated with reduced ED visits^{xxi}. CHCs are designed to promote wellness and serve populations that have traditionally faced barriers to accessing health services. TAIBU is one CHC in the GTA devoted to serving the black population that operates a SCD program one day per week where a haematologist sees patients with SCD. The haematologist prepared for this work by attending for 3 weeks in the GTA adult Centre of Excellence at the UHN, **and by regular communication/consultation with the Centre's Medical Director**. Patients receive evidence based care and there is a strong relationship between the CHC haematologist and the Centre of Excellence team. Patients with treatment challenges that cannot be met at the TAIBU are quickly referred to the UHN^{xxii}.

TAIBU is reaching out to two other CHCs in the GTA regarding the care of people with SCD^{xxiii}. These are examples of initiatives that can strengthen the education, social supports and overall management of SCD. CHCs are well positioned to improve the primary care of people with SCD and liaise with satellite centres and centres of excellence to support individualized care coordination, particularly for the small percentage of people with SCD who require more intensive clinical and social supports.

Advocacy groups

There are three advocacy groups, two provincial and one national that advocate for lifelong medical care and social supports for people with SCD in Ontario^{xxiv}

8.0 Membership

Sickle Cell Expert Panel				
LHIN	Member Name	Job Title and Organization	Chair/Co-Chair	Area of Expertise
1	Dr. Mark Awuku	Community Paediatrician, Windsor Associate Professor of Paediatrics, Department of Paediatrics, Schulich School of Medicine & Dentistry, Windsor Program, Western University		Physician
2	Ms Jill Craven	Director of Children's Care, Children's Hospital London Health Sciences Centre		Executive Administration
4	Dr. Uma Athale	Associate Professor, Department of Paediatrics, McMaster	-	Physician
4	Ms Danielle McKinlay	Information Controller, Decision Support Services McMaster Children's Hospital - Hamilton Health Sciences	-	Decision Support/ Health Information Management
4	Dr. Madeleine Verhovsek	Assistant Professor of Medicine, Division of Hematology and Thromboembolism, Consultant Laboratory Hematologist, Red Cell Disorders Laboratory, Hamilton Regional Laboratory Medicine Program, St. Joseph's Healthcare, Hamilton	Co-Chair	Physician
5	Dr. Sanjeev Luthra	Staff Pediatrician, Pediatric Sickle Cell Clinic, William Osler Health System		Physician
6	Ms Shauna Dowsley	Laboratory Manager, Hematology and Transfusion Medicine, Trillium Health Partners		Executive Administration
7	Dr. Isaac Odame	Haematologist, Division of Haematology/Oncology, SickKids. Professor, Department of Paediatrics, University of Toronto	Co-Chair	Physician
	Ms Jacqueline Flowers	Mother of child with SCD; involved with SCAGO		Patient Advocate
7	Ms Colleen Johnson	Nurse Practitioner, University Health Network, Red Blood Cell Disorders Program		Clinician
7	Dr. Carolyn Beck	Director of Inpatient Units, Division of Paediatric Medicine, SickKids		Physician
7	Ms. Marina Strzelecki	Clinical Pharmacist, SickKids		Clinician
7	Dr. Jennifer Stinson (Nurse Practitioner)	Mary Jo Haddad Nursing Chair in Child Health, Scientist, Child Health Evaluative Science; Associate Professor in the Faculty of Nursing, Institute of Medical Science and Institute of Health Policy Management and Evaluation at the University of Toronto, SickKids		Clinician

Sickle Cell Expert Panel				
LHIN	Member Name	Job Title and Organization	Chair/Co-Chair	Area of Expertise
7	Dr. Matthew Hodges	Emergency Physician, Mount Sinai Hospital		Physician
7	Dr. Dominick Shelton	Emergency Physician, Sunnybrook Health Sciences Centre		Physician
7	Christiane Gray-Schlehauf	Registered Nurse, Addiction Medicine and Chronic Pain, Centre for Addiction and Mental Health		Clinician
7	Dr. Richard Ward	Director, Red Blood Cell Disorders Program, Toronto General Hospital Associate Training Program Director, Adult Hematology Assistant Professor, University of Toronto		Physician
8	Dr. Olaniyi Ajisafe	Family Physician, North York, Ontario		Physician
9	Dr. Muhammad Akhter Hamid	Paediatrician, Rouge Valley Centenary Health Centre		Physician
9	Dr. Paks Madikiza	Paediatrician, Rouge Valley Centenary Health Centre		Physician
9	Dr. Larry Grossman	Haematologist, TAIBU Community Health Centre		Physician
11	Dr. Ewurabena Simpson	Assistant Professor, Division of Hematology/Oncology, Department of Pediatrics, Children's Hospital of Eastern Ontario		Physician
11	Ms. Fatiha Rochelle	Social Worker, Ottawa Hospital		Clinician
11	Dr. Ken Farion	Paediatric Emergency Medicine, CHEO		Physician

With, Mary Ellen Salenieks, Senior Project Manager, Provincial Council for Maternal and Child Health

Acute Pain Management for Sickle Cell Vaso-occlusive Crisis: Subgroup membership

Name	Role and Affiliation
Dr. Jennifer Stinson (subgroup chair)	Jennifer Stinson, RN-EC, PhD, CPNP Mary Jo Haddad Nursing Chair in Child Health Peter Lougheed CIHR New Investigator Scientist, Child Health Evaluative Sciences Nurse Practitioner, Chronic Pain Program The Hospital for Sick Children Associate Professor, Lawrence S. Bloomberg, Faculty of Nursing, University of Toronto Member, Expert Panel
Dr. Isaac Odame	Haematologist, Division of Haematology/Oncology, SickKids. Professor, Department of Paediatrics, University of Toronto Co-chair, Expert Panel
Dr. Madeleine Verhovsek	Assistant Professor of Medicine, Division of Hematology and Thromboembolism, Consultant Laboratory Hematologist, Red Cell Disorders Laboratory, Hamilton Regional Laboratory Medicine Program, St. Joseph's Healthcare, Hamilton Co-chair, Expert Panel
Ms. Lorraine Bird	Clinical Nurse Specialist, Acute Pain Management, SickKids
Ms. Salima Ladak	NP, Toronto General Hospital Acute Pain Service Coordinator, UHN Pain APN Committee
Dr. Larry Grossman	Community Hematologist, member QBP Expert Panel
Dr. Guy Petroz	Anaesthetist, Anesthesia and Pain Medicine, SickKids Assistant Professor, Department of Anaesthesia, University of Toronto
Ms. Mary Ellen Salenieks	Senior Project Manager, Provincial Council for Maternal and Child Health

Appendix 1: Screening for Sickle Cell Disease



NEWBORN SCREENING ONTARIO
DÉPISTAGE NÉONATAL ONTARIO

Screening for Sickle Cell Disease

Analysis by: Emeril Santander & Christine Davies (NSO)

For: PCMCH - SCD Quality Based Procedures project

Date: July 2015

Newborn screening for Sickle Cell Disease

Newborn Screening Ontario screens for 29 diseases including Sickle Cell Disease (SCD)

Screening for Sickle Cell Disease (SCD) in Ontario began November 24, 2006

Early detection permits improved health outcomes through early treatment and care

- Preventive antibiotic treatment can be started
- Education of family regarding medical management:
 - Fever, enlarged spleen, sickling crisis, aplastic crisis preventative measures
- Monitoring for stroke
- Avoids misdiagnosis



NEWBORN SCREENING ONTARIO
DÉPISTAGE NÉONATAL ONTARIO

Since 2006¹, 359 infants with Sickle Cell Disease have benefited from newborn screening

<u>Diagnosis</u>	<u>Count</u>
SS disease	210
SC disease	103
S-Beta thalassemia	40
HbS/HPFH	6
Grand Total	359

The birth prevalence in Ontario, Canada is 1:2814

** Incidence of new SCD diagnoses is likely slightly higher due to net immigration into Ontario. Dr. Ewurabena Simpson at CHEO has initiated a project that will help quantify this.*

¹ From 2006-2013 at Newborn Screening Ontario:
Total # of infants screened: 1,010,159



The Central and Central West LHINs have the highest proportion of newborns with Sickle Cell Disease or who are carriers

LHIN¹	Affected (2006-2013)			Carrier (2010-2014)		
	Count	Percent of Total	Rank	Count	Percent of Total	Rank
CENTRAL WEST (5)	81	23%	1	1229	17%	2
CENTRAL (8)	73	21%	2	1239	17%	1
CENTRAL EAST (9)	62	17%	3	1113	15%	3
TORONTO CENTRAL (7)	43	12%	4	616	9%	6
CHAMPLAIN (11) + GATINEAU	42	12%	5	658	8%	5
MISSISSAUGA HALTON (6)	26	7%	6	704	10%	4
HAMILTON NIAGARA HALDIMAND BRANT (4)	14	4%	7	426	6%	7
WATERLOO WELLINGTON (3)	< 6	1%	8	257	4%	8
SOUTH WEST (2)	< 6	1%	9	192	3%	9
ERIE ST CLAIR (1)	< 6	1%	10	143	2%	10
NORTH WEST (14)	< 6	0%	11	30	0%	13
SOUTH EAST (10)	< 6	0%	11	47	1%	12
NORTH SIMCOE (12)	< 6	0%	11	61	1%	11
NORTH EAST (13)	< 6	0%	11	61	1%	11
Grand Total	359	100%		6776	100%	

¹ LHIN was determined by the Forward Sortation Area (FSA) – first 3 digits of the maternal postal code submitted to NSO on the requisition form (i.e. maternal address at the time of birth)



Brampton is the community with the highest proportion of newborns with Sickle Cell Disease or who are carriers

Named Community ¹	Affected (2006-2013)			Carrier (2010-2014)		
	Count	% of Total	Rank	Count	% of Total	Rank
BRAMPTON	53	15%	1	855	13%	1
NORTH YORK	49	14%	2	775	11%	2
SCARBOROUGH	40	11%	3	670	10%	3
TORONTO	38	11%	4	391	6%	6
ETOBICOKE	28	8%	5	401	6%	5
OTTAWA/GATINEAU	25	7%	6	303	4%	7
MISSISSAUGA	24	7%	7	577	9%	4
YORK	13	4%	8	199	3%	8
AJAX	10	3%	9	167	2%	10
HAMILTON	7	2%	10	193	3%	9
NEPEAN	6	2%	11	92	1%	15
ORLEANS	< 6	1%	12	62	1%	20
PICKERING	< 6	1%	13	71	1%	19

¹ Named community was determined by the maternal postal code submitted to NSO on the requisition form and Canada Posts' community name associated with that postal code.



Toronto is the region in Ontario with the highest proportion of newborns with Sickle Cell Disease

Region ^{1,2}	Affected (2006-2013)			Carrier (2010-2014)		
	Count	Percent of Total	Rank	Count	Percent of Total	Rank
TORONTO	171	48%	1	2518	37%	1
PEEL	77	22%	2	1432	21%	2
OTTAWA/GATINEAU	42	12%	3	575	8%	3
DURHAM	19	5%	4	389	6%	5
YORK	11	3%	5	312	5%	6
HAMILTON	10	3%	6	231	3%	7
HALTON	7	2%	7	168	2%	9
OTHER	22	6%	9	1151	17%	3
Grand Total	359	100%		6776	100%	

¹ Region was determined by the maternal postal code submitted to NSO on the requisition form and the Association of Municipalities of Ontario's regions listing at www.amo.on.ca/YLG

² Regions with 5 or less affected individual/s are shown as 'Other'.



Limitations of NSO Data



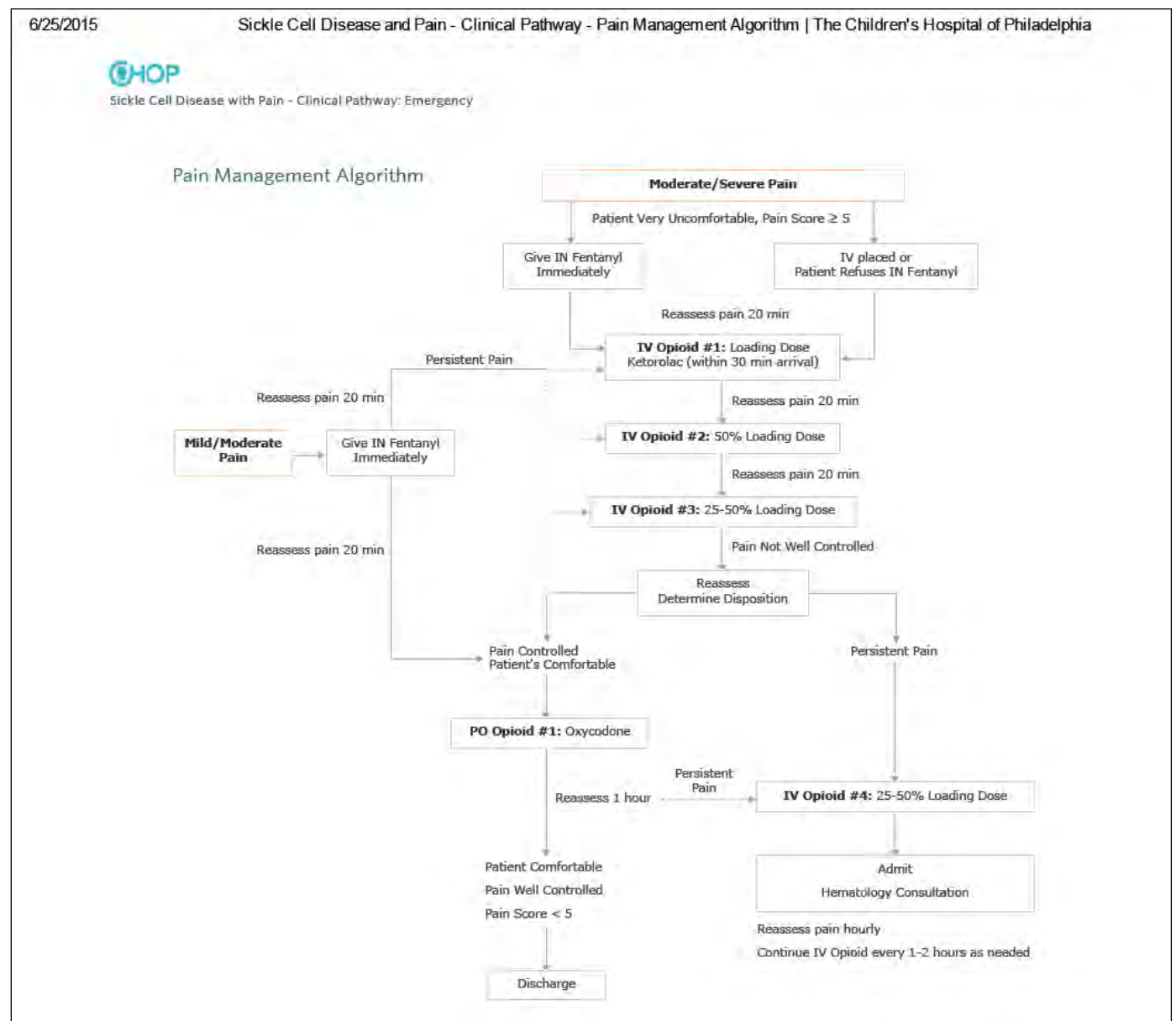
The data presented here only measures Ontario-born infants who had a newborn screen

The geography is calculated through self-reported maternal address at time of birth and does not necessarily reflect the current location of these infants

Appendix 2: ED visits and revisits, inpatient discharges and readmission for fiscal years 2011/12 and 2014/15

LHIN	Hospital	FY2011/12									FY2014/15						
		agegrp	ED Visit				Inpatient				ED Visit				Inpatient		
			ED Visits	Revisit ≤ 14 days	Revisit ≤ 30 days	Admis-sion rate	Inpatient Discharges	Readmit ≤ 14 days	Readmit ≤ 30 days		ED Visits	Revisit ≤ 14 days	Revisit ≤ 30 days	Admis-sion rate	Inpatient Discharges	Readmit ≤ 14 days	Readmit ≤ 30 days
7	(3969) HOSPITAL FOR SICK CHILDREN (THE)	adult	1	100%	100%	100%	2	50%	50%								
		peades	160	6%	14%	76%	141	5%	12%		157	6%	12%	84%	163	7%	14%
9	(3975) SCARBOROUGH HOSPITAL (THE)-SCAR.GEN.SITE	adult	82	68%	73%	17%	16	13%	13%		39	26%	41%	51%	21	10%	29%
		peades	1	0%	0%	0%	2	0%	0%		5	20%	20%	60%	7	14%	14%
11	(4085) OTTAWA HOSPITAL (THE)-GENERAL SITE	adult	107	36%	47%	50%	52	21%	29%		79	15%	29%	59%	59	14%	31%
		peades	1	0%	0%	100%	1	0%	0%		1	0%	0%	0%			
8	(4089) HUMBER RIVER REGIONAL HOSP-YORK-FINCH	adult	81	59%	67%	28%	23	13%	26%		40	15%	15%	53%	39	8%	8%
		peades	22	14%	14%	59%	19	0%	5%		10	10%	10%	40%	9	0%	0%
11	(4091) CHILDREN'S HOSPITAL OF EASTERN ONTARIO	peades	43	9%	14%	84%	31	3%	6%		38	5%	8%	68%	35	3%	6%
7	(4110) MOUNT SINAI HOSPITAL	adult	62	48%	63%	35%	21	19%	43%		24	71%	75%	29%	15	27%	27%
9	(4139) ROUGE VALLEY HEALTH SYSTEM-CENTENARY	adult	35	20%	29%	60%	25	8%	20%		36	11%	19%	69%	23	13%	22%
		peades	37	3%	8%	73%	43	19%	26%		33	9%	24%	85%	33	15%	27%
7	(4205) SUNNYBROOK HEALTH SCIENCES CENTRE	adult	32	50%	69%	72%	20	40%	60%		1	0%	100%	100%	3	0%	0%
		peades	44	55%	59%	27%	15	33%	40%		30	50%	63%	37%	13	0%	31%
7	(4209) TORONTO EAST GENERAL HOSPITAL (THE)	peades									1	0%	0%	100%	1		
8	(4233) NORTH YORK GENERAL HOSPITAL	adult	57	25%	33%	72%	42	19%	21%		37	16%	32%	73%	31	13%	16%
		peades	4	0%	0%	75%	4	0%	0%		1	0%	0%	0%			
5	(4245) WILLIAM OSLER HEALTH SYSTEM-ETOBICOKE	adult	59	42%	51%	58%	33	15%	27%		17	12%	12%	88%	16	6%	13%
		peades	10	0%	0%	100%	13	8%	15%		10	30%	30%	70%	9	22%	33%
7	(4260) HUMBER RIVER REGIONAL HOSP-HUMBER MEM	adult	50	22%	34%	32%	23	4%	13%		20	15%	25%	50%	17	12%	12%
		peades	4	0%	0%	50%	1	0%	0%		2	0%	0%	50%			
7	(4265) UNIVERSITY HEALTH NETWORK-GENERAL SITE	adult	181	47%	58%	34%	64	11%	17%		130	38%	43%	42%	76	9%	17%
		peades															
4	(4286) HAMILTON HEALTH SCIENCES CORP-MCMMASTER	peades	35	20%	46%	89%	29	21%	45%		32	25%	38%	97%	39	21%	41%
5	(4685) WILLIAM OSLER HEALTH SYSTEM-CIVIC SITE	adult	93	23%	31%	83%	88	22%	28%		66	12%	23%	88%	74	15%	23%
		peades	67	7%	9%	84%	71	8%	14%		34	12%	21%	88%	44	14%	18%
6	(4751) TRILLIUM HEALTH PARTNERS-CREDIT VALLEY	adult	106	42%	57%	38%	38	32%	34%		100	39%	45%	55%	65	20%	26%
		peades	15	13%	13%	93%	16	19%	19%		13	0%	0%	77%	18	6%	6%
6	(4756) TRILLIUM HEALTH PARTNERS-MISSISSAUGA S	adult	33	39%	42%	64%	26	23%	38%		23	9%	13%	52%	14	0%	7%
		peades					1	0%	0%		2	0%	0%	100%	2	0%	0%

Appendix 3: Algorithms from other jurisdictions





ED Pathway for Evaluation/Treatment of Children with Sickle Cell Disease and Pain

Goals and Metrics

Sickle Cell Disease Patient with Pain

Triage Criteria (Critical/Acute)

MD/CRNP/RN Rapid Assessment

IV Access
CBC with Differential
Reticulocyte Count
[History and Physical](#)

Consider Stroke Protocol:

Severe or Atypical HA
Altered Mental Status
Focal Neurologic Findings
New Seizure

Learn More

[Evidence-Based Management of Sickle Cell Disease, Expert Panel Report, 2014](#)

MILD / MODERATE PAIN

Minimal Pain Management at home

Oral Analgesia

Oxycodone
< 6 yrs 0.15 mg/kg up to 2.5 mg
6-12 yrs 0.2 mg/kg up to 5 mg
> 12 yr 0.2 mg/kg up to 10 mg
AND
Ibuprofen 10 mg/kg
(If no NSAIDS in preceding 6 hrs)
Reassess
0-60 minutes after po dose
! pain adequately controlled, discharge at 1 hour
! pain not adequately controlled, proceed to IV analgesia

Pain Management Algorithm

MODERATE / SEVERE PAIN

Aggressive Pain Management at home

IV/IN Analgesia

Fentanyl 2 mcg/kg administered with NAD
Consider as bridge if severe pain to ease IV access
Does not replace need for IV morphine
Morphine 0.1-0.15 mg/kg/dose IV (max 7 mg/dose)
Order 1st prn dose (50% of initial dose at the same time)
OR
Hydromorphone 0.015-0.02 mg/kg/dose IV (max 2 mg/dose)
Order 1st prn dose (50% of initial dose at the same time)
AND
Ketorolac 0.5 mg/kg/dose IV (max 30 mg/dose)
(If no NSAIDS in preceding 6 hours)
Reassess Every 20-30 minutes
Document pain score using age-appropriate scale
Monitor for side effects and complications

Titration of Opioid Therapy

Administer 25-50% of initial Morphine or Hydromorphone loading dose every 20-30 minutes as patient tolerates until pain adequately controlled

After 3rd PRN morphine or hydromorphone dose

Pain not adequately controlled - Admit to Hematology Service
Pain adequately controlled, or consider transition to PO, possible discharge

Hematology Consultation

Home

Patients with the following:
Pain relief continues for a minimum of 60 minutes following oral analgesia
Pain is relieved after 1 or 2 doses of IV analgesia
Relief is maintained on oral analgesic therapy
Absence of other complications of sickle cell disease

[Pain Management Algorithm](#)

[Discharge Plan](#)

HACU Admission

Patients with the following:
≥ 6 months of age
Non-toxic
Uncomplicated vaso-occlusive pain event requiring multiple IV doses of analgesics

Inpatient Admission Criteria

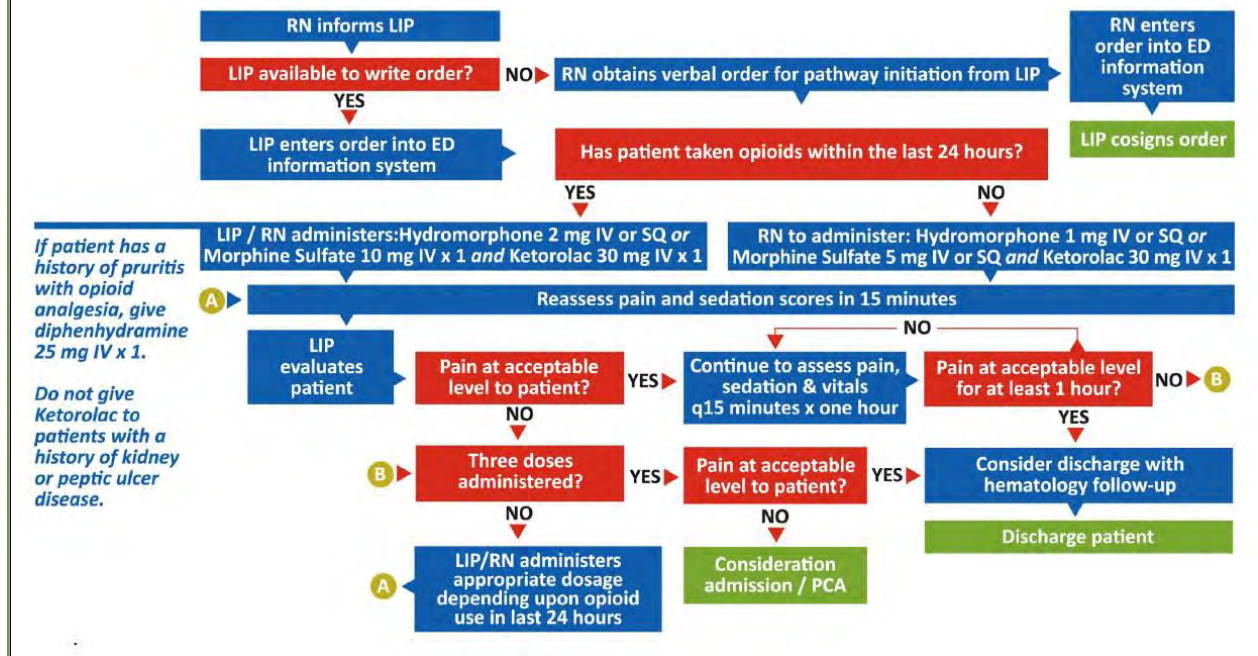
Patients with the following:
< 6 months of age
Focal neurologic findings
Acute chest syndrome
Splenic sequestration
Fever
Reticulocyte count < 1 % (unless Hgb > 10 g/dL)
Severe anemia (Hgb < 5 g/dL)

Posted: July, 2010

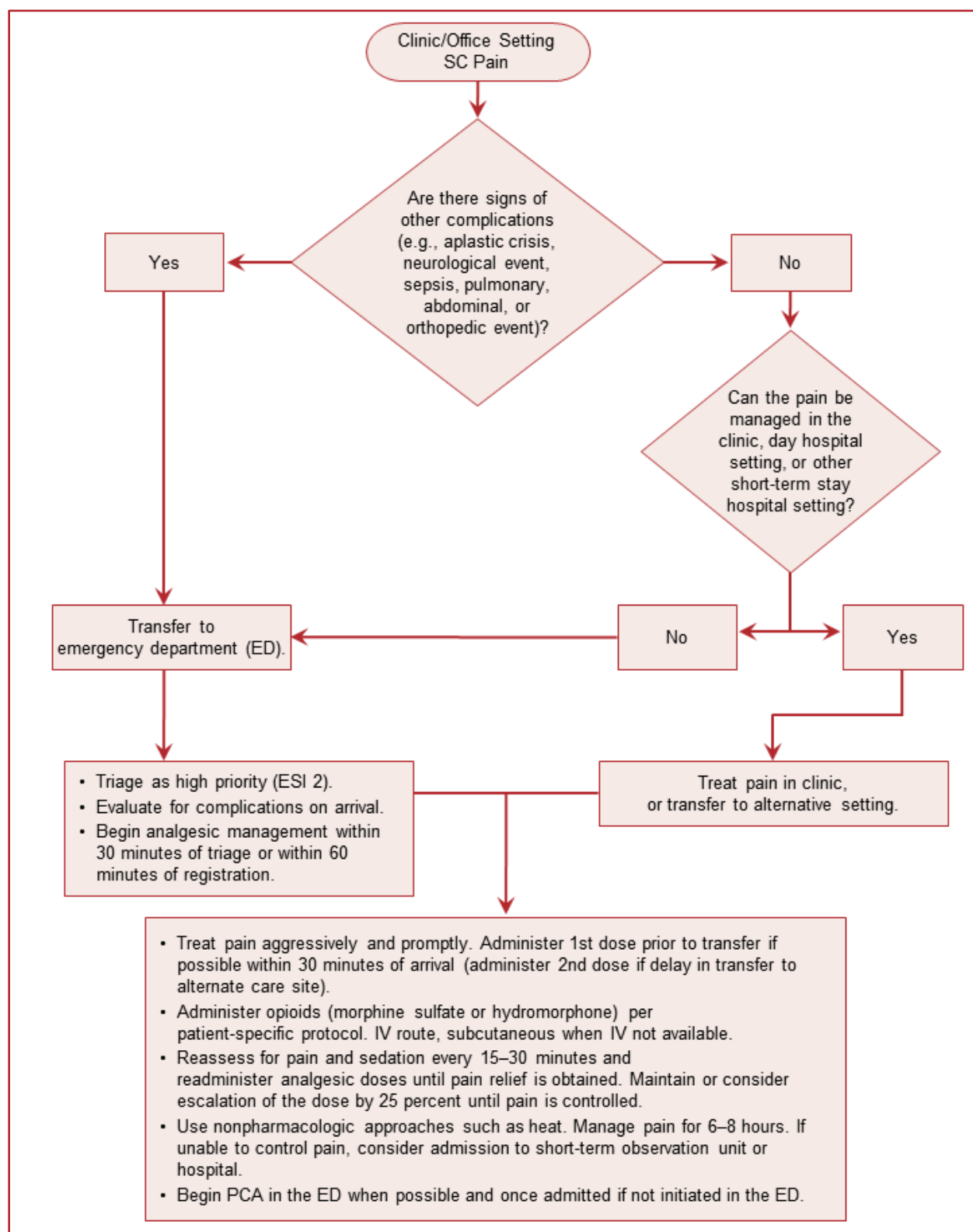
Revised: October 2011, October 2014

Authors: A. Ellison, MD, Lisa Payne, RN, C. Norris, MD, K. Smith-Whitley, MD, C. Jacobstain, MD, J. Lavelle, MD

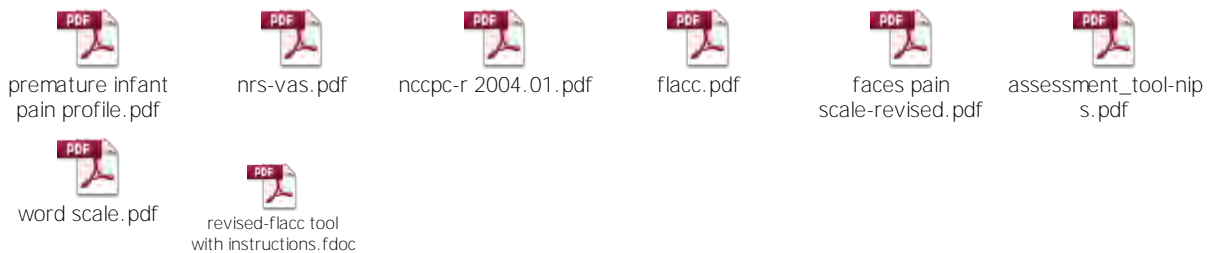
ED SCD Pain Management Algorithm



<http://www.ctscts.org/seed.html>



Appendix 4: Pain Scoring Tools



Appendix 5: Implementation of a Community Hospital-Based Fast Track Pathway for the Treatment of Acute Pain Episodes in Adults with Sickle Cell Disease

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Background: A growing number of adult patients with sickle cell disease (SCD) receive care in a community setting and often lack access to physicians with sickle cell expertise. To address this healthcare disparity we are testing a co-management medical home model with Hematology/Oncology (H/O) and Family Medicine (FM) physicians to facilitate evidence-based acute and chronic care. An Emergency Department (ED) Observation Unit based pathway for treatment of sickle cell pain developed at an academic medical center with a Comprehensive Sickle Cell Center (CSCC) has been adopted and modified to fit the needs of a community multi-specialty hospital with an unopposed FM residency program. The hospital serves a large sickle cell population in a predominantly rural setting with the closest CSCC 180 miles away.

Methods: Pathway development was facilitated by having a formal meeting for the community hospital physicians and staff at the academic medical center and sickle cell experts providing ongoing on site consultation at the community hospital. Protocols for the community hospital were produced with input from physicians, nurses, advanced practice providers, and support services at multiple meetings. Adult patients with SCD presenting to the ED with pain are triaged at Emergency Severity Index Level 2 for evaluation by the ED physician. The ED protocol uses specific criteria to identify patients with uncomplicated pain. Patients presenting with abnormal vital signs (other than mild tachycardia), fever, pregnancy, or apparent other sickle cell-related complications are excluded.

Patients qualifying for the pathway are directly admitted to the SCD unit (a hospital room with 4 infusion chairs on the H/O floor exclusively designated for care of sickle cell patients). Following intake evaluation by the nurse, a clinician is notified to evaluate the patient and provide orders for intravenous fluids and opioid patient controlled analgesia (PCA) which is administered according to hospital guidelines. PCA by the subcutaneous route is used if intravenous access is not readily available. A CBC is obtained whereas other laboratory testing and imaging studies are ordered based on clinical indications. H/O physicians and nurse practitioners cover the unit weekdays 8:00am-5:00pm and FM residents cover nights and weekends with back up by the on call H/O physician. Patients can be treated in the SCD unit up to 23 hours. For patients discharged home a follow up phone call by an H/O nurse will be placed within 3 days and an outpatient clinic appointment is scheduled to be within 7 days. Monthly quality assurance meetings are attended by H/O, FM, and ED physicians as well as nursing, pharmacy and administrative staff from the ED and H/O inpatient service to review process issues and patient outcomes. Consultation is provided by academic physicians with sickle cell expertise (H/O and ED) who attend each meeting in person or by conference call.

Results: From March 5-June 30, 2014, 67 patients accounted for 271 visits to the SCD unit. The mean time in the unit was 13.6 hours. The mean pain score on admission was 8.7/10 and reduced to 4.9 upon discharge. PCA drug, pump setting, and dosage are recorded to be used for future visits. Over the 4 months 91.1% of the patients were discharged home from the unit. Six patients accounted for 31% (84) of the visits with only 4 hospital admissions.

Conclusions: A fast track pathway for the treatment of acute sickle cell pain coordinated between ED, H/O, and FM physicians has been implemented at a community hospital using an Observation Unit based treatment program. During the entire initial experience the majority of patients have been discharged home with adequate pain relief. In the future the impact of the program will be evaluated including effect on frequency of hospitalizations, outpatient follow up, patient satisfaction, and cost effectiveness. The pathway can be adapted to other community hospital settings where sickle cell expertise is not locally available.

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