



Guideline for Disease Management in Correctional Settings

ADOLESCENT SICKLE CELL DISEASE

Recommended Resources to Support Evidence-Based Practice and Quality Improvement

NCCHC issues guidelines to assist correctional health care clinicians in evidence-based decision making. For specific clinical practice guidelines and recommendations, please see the resources listed on page 3.

Introduction

Although clinical guidelines are important decision support for evidence-based practice, to leverage the potential of guidelines to improve patient outcomes and resource use, NCCHC recommends that health care delivery systems also have components including primary care teams, other decision support at the point of care (such as reminders), disease registries, and patient self-management support. These components have been shown to improve outcomes for patients with chronic conditions. In addition, we recommend establishment of a strategic quality management program that supports ongoing evaluation and improvement activities focused on a set of measures that emphasize outcomes as well as process and practice. For information on the chronic care model, model for improvement, and outcomes measures, see the resources listed on page 3.

Sickle Cell Disease Care in Corrections

The care of sickle cell disease in corrections is organized into four components:

- Assessment and monitoring of disease management and control
- Treatment with medications or therapies to prevent crises
- Prompt and appropriate treatment of crises
- Education of patient and correctional staff about the disease process and methods to prevent and treat crises

Sickle cell disease is a genetic disorder that results in structurally abnormal hemoglobin that alters the flexibility of red cells and causes painful crises, significant anemia, and many serious complications. Persons who are most likely affected are homozygous for hemoglobin S (SS disease) or have a combination of hemoglobin S and hemoglobin C (SC disease). Persons who are heterozygous for hemoglobin S do not have sickle cell disease but may have some complications. Patients with sickle cell disease can develop four types of crises: aplastic crisis, sequestration of red cells in the spleen, hyper-hemolytic crises, and the most common, vaso-occlusive crises. These may occur at any time and can result in permanent organ damage.

At the age when a youth enters a correctional facility, the diagnosis of sickle cell disease is likely to be known. In some cases, the youth may not distinguish between sickle cell trait and disease. If the clinical history is not sufficient to separate the two, it will be necessary to perform laboratory testing. Youth with known or suspected sickle cell disease require immediate inquiry into their health status at the time of receiving screening. The following current complaints should be evaluated urgently by a physician: fever, painful crisis, abdominal pain, dyspnea, central nervous system symptoms (weakness, change in sensation, problems speaking, recent seizure), priapism (prolonged erections), and new onset problems with vision. The health assessment requires a complete history of the disease and complications as well as a psychosocial history and total lifetime transfusion history. Records of previous and ongoing care are necessary. The physical examination should include a body mass index, Tanner staging, blood pressure, and funduscopic, lung, cardiac, and neurological examination.

The clinician and patient will focus on reducing acute manifestations of the disease such as painful crises, anemia crises, acute chest syndrome, stroke, priapism, and infections. Long-term management aims at preventing or reducing the complications. Immunizations may be required. Consultation with a specialist in sickle cell disease will help guide preventive care and acute management of intercurrent illness. Patients who previously suffered or developed a serious complication while in custody must see a sickle cell specialist. Patients in a crisis situation are likely to need narcotics, and clinicians and custody staff should be prepared for this.

Definitions of clinical severity:

- Mild: Infrequent pain or anemic crises once or less every 2 years and no evidence of current organ damage.
- Moderate: History of one or two pain or anemic crises per year and/or only mild and stable organ dysfunction. No obvious strokes in past.
- Severe: History of three or more pain or anemic crises per year, or chest syndrome and/or priapism with continuously deteriorating organ function, or evidence of a stroke in the past. Most youth with sickle cell or sickle thalassemia will be considered severely affected.

Improving control of sickle cell symptoms involves the use of multiple modalities: adequate immediate pain control, medications and other treatments, avoiding dehydration, and periodic screening for organ damage.

Definitions of control:

- Good control: No pain crises since last visit (last 6 months) or the need for only nonsteroidal anti-inflammatory drugs and normal vital signs, no emergency department visits or hospitalizations, and stable organ function and normal central nervous system function by clinical examination.
- Fair control: One to two pain or anemic crises over the past 6 months requiring management in an emergency department or hospitalization, or an infection requiring emergency department management or hospitalization, or abnormal (but stable) renal and liver function, or central nervous function abnormalities, but no evidence of acute change such as a new stroke.
- Poor control: Three or more pain or anemic crises requiring emergency department management or hospitalization in last 6 months or worsening organ function tests, or evidence of acute central nervous changes such as a stroke.

Sickle cell disease is frustrating for both the patient and clinician in any setting. However, during incarceration the limit in freedom to seek immediate care for episodes of pain and the need for multiple preventive care consultations adds significant stress for the patient. The patient also lacks social and other distractions that help in coping with this painful, debilitating disease.

Clinicians' limited knowledge of sickle cell disease, inadequate assessment of pain, and biases against opioid use are barriers to effective pain management. The biases are based on lack of knowledge about opioid tolerance and physical dependence and confusion with addiction. Unwarranted fear of addiction is common and may be heightened in correctional settings. The patient's experience of pain should guide the assessment of pain. Failure to rapidly and effectively address pain causes unnecessary stress and suffering and often prolongs pain episodes or leads to chronic pain. Custody staff who work with sickle cell patients will require training to understand the need for narcotics in these patients. A comprehensive psychosocial evaluation should be performed yearly or more often for patients with frequent pain.

Custody staff who work with sickle cell patients will require training to recognize signs of crises, including priapism, and to prevent crises by assuring adequate hydration. Very small fluctuations in hydration can have profound effects on sickling. Many patients, even some with milder variations, including carriers of sickle cell, are unable to concentrate their urine. Thus, it is important to realize that during hot weather, strenuous exercise, or fever, they may become dehydrated and require parenteral fluids.

Priapism may be misunderstood by staff and ignored or even ridiculed by custody staff. All staff working with sickle cell patients need to know that priapism is not self-induced and is a medical emergency.

Many of the crisis situations must be emergently addressed and require rapid transfer to emergency departments, preferably with sickle cell expertise or available consultants. If the facility houses sickle cell patients, staff at all levels should be alert to the need for rapid response. The logistics of these transfers can be complex and time consuming so it is best to make advance plans with the custody staff so that they understand the urgency and will be prepared to respond rapidly.

Sickle cell patients have no outward stigmata of severe disease yet many have reduced work tolerance because of anemia or previous neurological insult. A substantial number have neurocognitive problems, so all patients should be tested for these defects. More than 50% of sickle cell patients who have had an overt or silent stroke will have decreased academic ability and neuropsychological problems and may eventually require custodial care. If neurocognitive defects are found, appropriate remedial plans should be offered to the patient. In addition, a full workup of risk for stroke is indicated.

These patients will incur significant costs for medical care. Many of the required screening tests are likely to identify risks such as risk of stroke, eye disease, renal disease, and hepatobiliary disease. It is expensive and time consuming to assess these risks and treat them if present. Because of the relative rarity of sickle cell disease and its many complications, knowledgeable specialists are necessary to guide treatment. This usually means many trips outside the facility. The administrator and medical staff should meet to discuss the implications when a sickle cell patient first arrives. Forewarning and understanding of sickle cell disease will greatly improve the staff's ability to accommodate the needs of these patients.

Quality Improvement Measures

The following quality improvement measures are suggested, but they are not intended to be a complete list necessary to ensure a successful sickle cell disease management program in a juvenile correctional setting. We recommend that the improvement measures for a patient population be reported at a facility level and at a provider or team level. These indicators should be compared over time to correlate improvement.

- Percentage of patient charts with initial history and physical recommended elements
- Percentage of sickle cell patient charts with annual eye examinations
- Percentage of sickle cell patients with renal function tests
- Percentage of sickle cell patients who receive prompt and adequate treatment during acute pain crises
- Percentage of sickle cell patients whose management is guided by an expert in sickle cell disease, at minimum a hematologist

Recommended Resources to Support Evidence-Based Practice and Quality Improvement

RESOURCE	The Management of Sickle Cell Disease (2002)
SOURCE	Division of Blood Diseases and Resources, National Heart, Lung, and Blood Institute, National Institutes of Health
URL	http://www.nhlbi.nih.gov/health/prof/blood/sickle/index.htm
RESOURCE	Chronic Care Model: Meet the Needs of Specific Populations
SOURCE	Based on the model developed by Ed Wagner MD, MPH, MacColl Institute for Healthcare Innovation, Group Health Cooperative of Puget Sound, and the Improving Chronic Illness Care program. Available from the Institute for Healthcare Improvement
URL	http://www.ihl.org/knowledge/Pages/Changes/MeettheNeedsofSpecificPopulations.aspx

RESOURCE How to Improve / Model for Improvement
SOURCE Associates in Process Improvement. Available from the Institute for Healthcare Improvement
URL <http://www.ihl.org/IHI/Topics/Improvement/ImprovementMethods/HowToImprove>

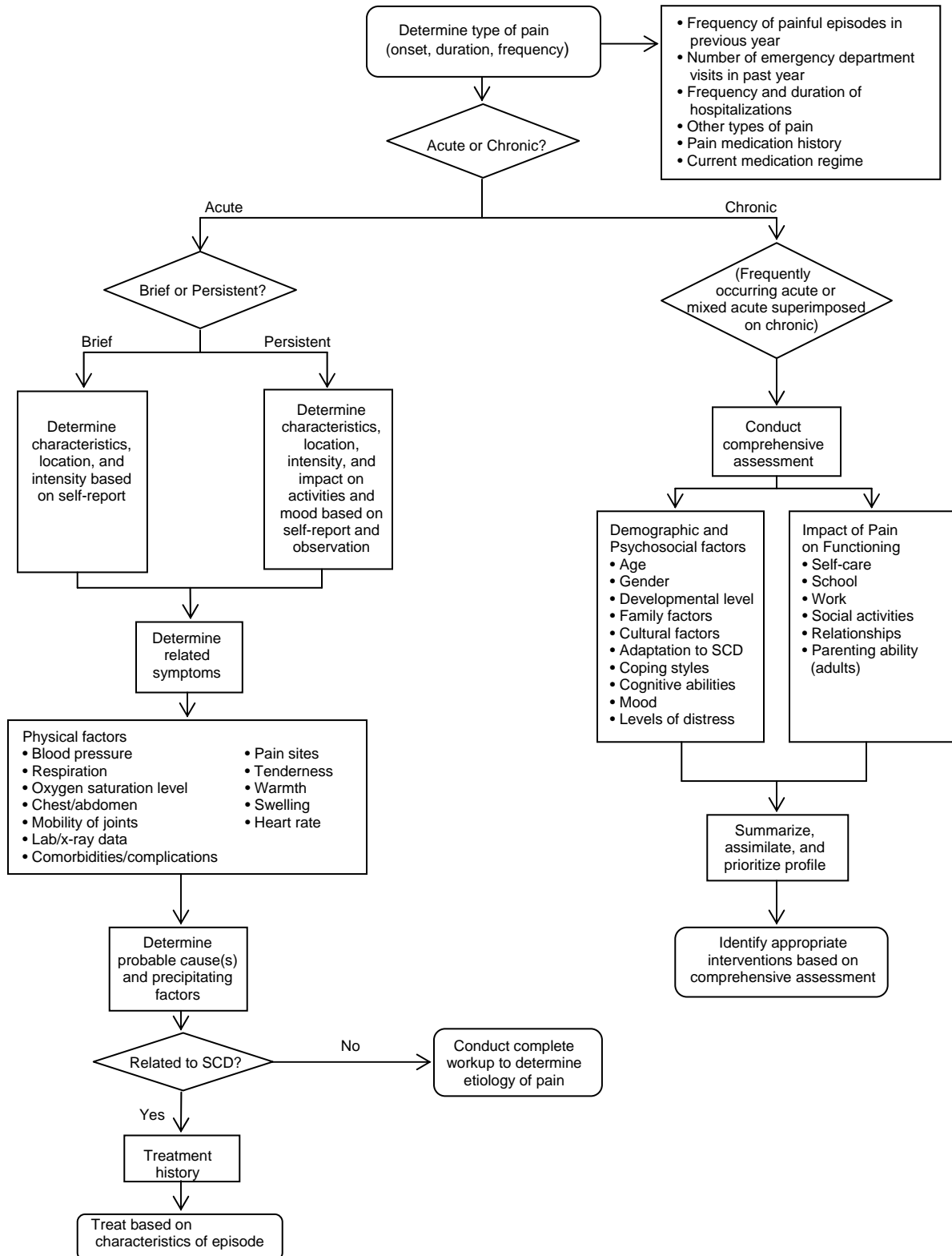
RESOURCE Measures
SOURCE Institute for Healthcare Improvement
URL <http://www.ihl.org/knowledge/Pages/Measures/default.aspx>

RESOURCE HEDIS & Quality Measurement
SOURCE National Committee for Quality Assurance
URL <http://www.ncqa.org/tabid/59/Default.aspx>

Last reviewed: October 2011
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For the latest version, go to
<http://www.nchc.org/resources/clinicalguides.html>

Appendix 1

Assessment Overview (From *The Management of Sickle Cell Disease*, NHLBI, 2002)



Appendix 2

Suggested Management for Adolescent Sickle Cell Disease in Correctional Settings

Admission		
Identification	History	Physical Examination
<ul style="list-style-type: none"> - Determine type through inquiry of fever, painful crisis, abdominal pain, dyspnea, CNS symptoms (weakness, change in sensation, problems speaking, recent seizure), priapism, and new onset problems with vision - Records from previous clinicians - Interview youth's parents 	<ul style="list-style-type: none"> - Sickle cell trait will not have a history of painful episodes - Complete history regarding clinical events: variety, frequency, and length of crises, along with treatments, to help predict future events - Hospitalizations, both inpatient and ICU, including length of stay, for insight into severity and specific areas of vulnerability - Obtain a total lifetime transfusion load to predict possible iron overload - Inquiry into erectile dysfunction 	<ul style="list-style-type: none"> - Body mass index (BMI) - Vital signs and blood pressure - Tanner staging - Funduscopic examination - Cranial nerves - Lymphadenopathy - Lung - Cardiovascular exam - A palpable spleen likely indicates a variant of sickle cell disease - Thorough neurologic exam - Signs of previous ischemia to extremities - Genital exam
Diagnostics	Special	Treatment
<ul style="list-style-type: none"> - CBC - Reticulocyte count - Peripheral smear - Hemoglobin electrophoresis - Quantitative measurement of hemoglobins A, S, A₂, and fetal - Liver function panel - Kidney tests - Lipid profile - Urinalysis - Microalbuminuria - Hepatitis B and C testing for previous blood transfusions 	<ul style="list-style-type: none"> - Screening for high blood pressure, which may signal kidney disease or other disorders - Psychosocial history, including coping style, family engagement, depression, and psychiatric hospitalizations 	<ul style="list-style-type: none"> - Consultation with sickle cell disease specialist - Hydroxyurea should be prescribed and monitored by a SS specialist - Painful events are the most common cause of emergency room visits and hospitalizations; painful crises often require the use of opioids and affect the patient's ability to cope
Complications		
Infections	Splenic Sequestration	Acute Chest Syndrome
<ul style="list-style-type: none"> - Provide appropriate immunizations - 23-valent pneumococcal polysaccharide, haemophilus influenza type B, meningococcal, and hepatitis B vaccines - Annual influenza vaccination and revaccination for pneumococcus after 5 years is recommended - Higher risk for bacterial infections so a SS youth with a fever of 38.5° C or higher should be evaluated for sepsis in a facility equipped to provide rapid resuscitation until it is clear the patient is not septic - Many patients with chest pain are infected with atypical organisms 	<p>Splenic sequestration is defined as a fall of at least 2 g/dL from steady-state hemoglobin concentration, evidence of increased erythropoiesis with a markedly increased reticulocyte count, and a rapidly enlarging spleen</p>	<p>The association of a new pulmonary infiltrate during an episode of acute chest pain and respiratory symptoms is a major cause of mortality in patients age 20 years or older</p> <ul style="list-style-type: none"> - Fat emboli - Manage these patients in a hospital regardless of age - Oxygen should be begun during transport to the hospital
Cardiovascular	Ophthalmologic	Other
<ul style="list-style-type: none"> - Cardiohypertrophy is common - Hyperactive precordium - Systolic murmurs - Physical work capacity is reduced in 50% of adults and in 60% to 70% of children and is related to the degree of anemia 	<ul style="list-style-type: none"> SS vaso-occlusive events can affect all vascular levels in the eye - Annual ophthalmological exams are necessary 	<ul style="list-style-type: none"> - Priapism can lead to permanent damage to the erectile function - Intrauterine devices should be avoided - The incidence of overt stroke, silent stroke, and transient ischemic attack is 40% by 20 years of age