Splenic Sequestration:

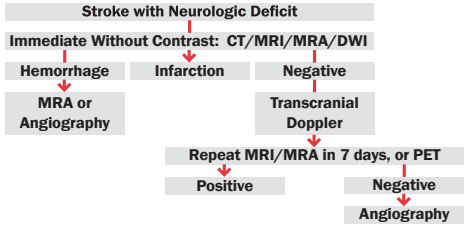
Acute splenomegaly, pallor, tachycardia, or lethargy can be the first clinical signs of a potentially life-threatening splenic sequestration crisis. The child's hemoglobin may drop acutely (to as low as 1-3 g/dL), resulting in hypovolemic shock and death within hours of initial onset. Prompt treatment with volume expanders and cautious blood transfusion in aliquots to reverse the hypovolemic shock may be required.

Acute Chest Syndrome:

Characterized by chest pain, cough, fever, hypoxia, and lung infiltrates. Acute chest syndrome may be the result of sickling in the microvasculature causing pulmonary infarction/emboli or pneumonia. Pleuritic chest pain is the most common presenting complaint in adults. Fever, cough, tachypnea, hypoxemia, or abdominal pain are common presentations for infants and children. It is always best to assume an underlying infectious etiology. Treatment with ceftriaxone and azithromycin is indicated. Chest x-ray findings may lag behind clinical symptoms. Treatment with exchange transfusion should be used for patients who have multi-lobar involvement, hypoxemia, or progressive respiratory distress. Simple transfusion may be used if the baseline hemoglobin is low.

Stroke Evaluation and Emergent Intervention:

Signs and symptoms of stroke include: seizures, acute severe headache, slurred speech or aphasia, somnolence or disorientation, weakness or numbness - usually on one side of the body, painless limp, visual or auditory changes. Rapid evaluation and monitoring of progression of symptoms (i.e. increased intracranial pressure) are crucial. Hyperventilation therapy should be avoided. Cerebral edema should be managed pharmacologically. Mechanical ventilation may be necessary. Seizures are common and require anticonvulsant therapy. Transfusion of normal red blood cells emergently will help prevent the progression of the acute stroke. Partial exchange transfusion or a 1-volume exchange transfusion should be used to decrease the level of HbS to ≤ 30%. Simple transfusion (transfusing PRBC without prior or concomitant removal of the patient's blood) is generally not recommended in this situation. Thrombolytic (TPA) therapy is not indicated in sickle cell related stroke.



FOR MORE INFORMATION, CONTACT:

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317-233-1357 / Toll Free: 1-888-815-0006

Martin Center Sickle Cell Initiative
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SCACURE Networks, Inc.
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North Central Indiana Sickle Cell Initiative
South Bend, IN
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Indiana Sickle Cell Consortium (ISCC)
www.indianasicklecell.org

EMERGENCY MANAGEMENT

of Sickle Cell Patients

Care of Patients with Sickle Cell Disease for Hospital and Emergency Room Personnel

Indiana Hemophilia & Thrombosis Center

1 **BELIEVE ME** WHEN I SAY I HAVE PAIN.

2 TREAT ME **PROMPTLY.**

3 **CALL MY HEMATOLOGIST** FOR TREATMENT ADVICE.

4 FOR SERIOUS SYMPTOMS, **TAKE ACTION.**

PHYSICAL OR RADIOGRAPHIC FINDINGS ARE ONLY PRESENT IN ABOUT 50% OF ACUTE VASO-OCCLUSIVE PAINFUL EPISODES

PAINFUL VASO-OCCLUSIVE EPISODE:

- **Dactylitis** (painful swelling of the hands or feet) is often the first manifestation of sickle cell disease seen in affected infants
- In older children and adults, **musculoskeletal pain** is the most common complaint. It may be difficult to distinguish a vaso-occlusive pain episode from osteomyelitis, septic arthritis, toxic synovitis, rheumatic fever, or gout.
- For patients with **abdominal complaints**, pancreatitis, cholecystitis, urinary tract infection, pelvic inflammatory disease, ectopic pregnancy, pneumonia, or malignancy must be ruled out

Assessment

- ***Believe the patient***
- History and physical examination, including prior experience with pain management agents
- Triage as High priority (Emergency Severity Index [ESI]2)
- Document location and intensity of pain on a simple measurement scale
- Note that symptoms of opioid withdrawal can mimic a pain crisis (hyperalgesia)
- Vital signs with oxygen saturation measurement
- CBC with differential, reticulocyte count, CMP
- Blood cultures for fever $\geq 101^{\circ}$ F
- Serum or urine pregnancy test for pre-menopausal women
- Radiographs as indicated

Action

- A delay in treatment of fever, acute chest syndrome, stroke, or splenic sequestration can result in increased morbidity and mortality
- Use the patient's written treatment plan if available and refer to the tables at right if no treatment plan is available.
- **AVOID** use of meperidine (increased risk of seizure in sickle cell)
- Synthetic opioids (pentazocine, butorphanol, nalbuphine) should be avoided because of antagonist induction of withdrawal symptoms or psychomimetic effects
- Use acetaminophen and ibuprofen (or IV ketorolac) as adjunctives to opioid therapy
- Hydrate with D5-0.5NS at maintenance rate. Consider a rate of 1.5 times maintenance if acute chest syndrome is **not** suspected
- Oxygen is indicated only if O_2 saturation is $\leq$95%
- Administer an oral or parenteral analgesic agent mutually agreed upon with the patient
- Contact the patient's hematologist for treatment recommendations

Assessment of Efficacy

- ***Believe the patient***
- Monitor for side effects of opioid analgesia: respiratory depression, nausea, vomiting, pruritus, hypotension, secretion of antiduretic hormone, urinary retention, or changes in seizure threshold. Consider use of oral diphenhydramine and ondansetron for supportive care if indicated
- A respiratory rate $\leq$ 10/minute is a sign of opioid induced respiratory depression
- 15 – 30 minutes after initial administration of IV analgesia, reassess pain location and intensity
- If no relief after 15 minutes, 50% of initial opioid dose should be repeated. If the patient is mildly sedated but still reporting pain, 25% of the initial dose should be given
- If the patient is comfortable for 3 hours, administer an oral narcotic and observe for another hour. If pain returns within 30 minutes, repeat the initial IV dose. If pain persists, admit and initiate a PCA infusion

EXAMPLES OF ANALGESICS AND SUGGESTED DOSING:

(opioid dose should be chosen based upon prior opioid exposure, pain history, and clinical presentation)

Severe Pain: *IV* *opiates for severe pain*

Drug	Children (<50 kg)	Adults	Opiate-tolerant adults (>100 morphine equivalents / day)
Morphine (IV)	0.1-0.2 mg/kg IV every 3 hours (max dose 10 mg)	5-7.5 mg IV q 30 mins	7.5-10 mg IV q 30 mins
Hydromorphone (IV)	0.01-0.02 mg/kg IV every 4 hours (max dose 2 mg)	2 mg IV q 30 mins	2-4 mg IV q 30 mins
Fentanyl (IV)	1-2 mcg/kg IV every 2 hours	50 mcg IV q15 min PRN	75-100 mcg IV q15 min PRN

Moderate Pain: *Oral* *opiates for moderate pain*

Drug	Children (<50 kg)	Adults
Oxycodone (oral)	0.1-0.2 mg/kg PO every 4 hours	5-10 mg PO every 4 hours
Hydrocodone- acetaminophen (oral)	Hydrocodone 0.1-0.2 mg/kg PO every 4 hours	Hydrocodone 5-10 mg PO every 4 hours
Morphine (oral)	0.2 - 0.3 mg/kg PO every 4 hours	15-30 mg PO every 4 hours
Hydromorphone (oral)	0.03-0.08 mg/kg PO every 4 hours	2-8 mg PO every 4 hours

PCA: (suggested starting dose; titrate to effect)

Drug	Children (<50 kg)	Adults
Morphine	0.01 mg/kg/hour basal with 0.01 mg/kg demand every 15 minutes PRN	1.5 mg/hour with 1.5 mg demand every 15 minutes PRN
Hydromorphone	0.004 mg/kg/hour basal with 0.004 mg/kg demand every 15 minutes PRN (max. 0.2 mg/hour)	0.2 mg/hr with 0.2 mg demand every 15 minutes PRN

If PCA demands are excessive, increase the continuous infusion rate by 50% and observe for relief.
Initiate bowel regimen with any extended course of opioids.

FEVER EVALUATION AND MANAGEMENT:

The most common causes of infections in sickle cell disease are (in order of frequency): Streptococcus pneumoniae, Hemophilus influenzae, Neisseria, Salmonella, Mycoplasma, Staphyloccus aureus, Escherichia coli, and Streptococcus pyogenes.

Infections cause more morbidity, disseminate more rapidly, and are more difficult to eradicate in persons with sickle cell disease. Infections can precipitate aplastic crisis and exacerbate hemolytic events and can also precipitate vaso-occlusive episodes.

All sickle cell patients $\geq$3 months of age with fever (T>101°F) should have blood cultures collected and should receive IV or IM ceftriaxone 50-75mg/kg. Give adult dose 1-2g. Additional workup and treatment should be dictated by history and clinical exam.

All infants 12 months of age and younger with SCD and fever should be admitted to the hospital for observation and IV antibiotics until blood cultures are negative X 48 hours.

Fever in infants <3 months of age should be managed per standard insitutional protocol.

*If you have any questions about a patient's care, please contact the IHTC **immediately** at **317-871-0000***