Louisiana Medicaid | Provider UPDATE

Volume 33, Issue 15 | November 2017

Sickle Cell Disease

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BACKGROUND

Sickle cell disease (SCD) is a life-threatening global health issue that affects approximately 100,000 Americans, with an additional three million that carry the sickle cell trait (SCT). The disease occurs in approximately one out of every 365 Black or African American births and one out of every 16,300 Hispanic-American births. Approximately one in every 13 Black or African-American babies are born with the SCT.[1]

SCD is an inherited red blood cell (RBC) disorder characterized by sickle hemoglobin (Hgb S) and rigid "sickle" shaped RBCs. Normal RBCs have a lifespan of 90 to120 days, whereas the life span of sickled RBCs is 10 to 20 days resulting in anemia and fatigue. The sickled RBCs are unable to carry oxygen efficiently and tend to adhere together, which causes blocked vessels and a variety of complications.[2]

COMPLICATIONS

Vascular occlusion, the hallmark of SCD, causes tissue ischemia and oxygen deprivation. This can lead to a vaso-occlusive crisis (VOC), or acute pain crisis, which is the most common acute complication of SCD. These crises can last for days or weeks and typically occur in the lower back, legs, hips, abdomen and chest. Acute chest syndrome (ACS), another acute complication of SCD, involves pain in the chest and evidence of pulmonary infection. ACS can be life-threatening and is the leading cause of mortality in SCD patients.[2] In addition to VOCs and ACS, other common acute complications are acute stroke, acute anemia, fever associated with infection, hepatobiliary manifestations, acute kidney injury, and splenic sequestration. Chronic pain, avascular necrosis, leg ulcers, pulmonary hypertension, renal impairment, stuttering/recurrent priapism and ophthalmologic complications are the most common chronic complications associated with SCD.[3] See Table 1 for more information regarding complications of SCD.

INFECTION RISK

The spleen of an SCD patient becomes fibrotic and reduces in size due to repetitive infarctions, which results in functional asplenia. Patients with functional asplenia are at serious risk of infectious diseases and should take extra precautions, such as immunizations, antibiotic prophylaxis when indicated, and medical attention with fever greater than 101.5°F.[4,5]

NON-DRUG TREATMENT

By providing normal RBCs via blood transfusions, the risk of life-threatening complications is decreased for SCD patients. Stroke, acute chest syndrome and symptomatic anemia are acute indications for blood transfusions. Regularly scheduled transfusions can help reduce the risk of stroke, acute chest syndrome, painful events, priapism, and pulmonary hypertension in SCD patients.[4] Refer to the 2014 Evidence-Based Report on the Management of Sickle Cell Disease for detailed recommendations on the use of blood transfusions.[6] Hemapoietic cell transplantation (HCT) has been used in only a small sample of patients although studies have demonstrated the procedure to be curative in certain patients.[4]

DRUG TREATMENT

Primary drug therapies used in SCD: [4,6]

- Antibiotics and Immunizations to reduce infection risk
- Analgesics to control pain
- Hydroxyurea to prevent or reduce acute and chronic complications
- Chelation therapy to manage blood transfusion-related iron overload

Antibiotics and Immunizations

Severe infections are a major cause of death in patients with SCD, particularly in children younger than five years of age. Bacteremia, meningitis, and pulmonary infections can occur and are commonly due to organisms such as *S. pneumoniae, H. influenzae, N. meningitidis and Salmonella spp*. as well as atypical infections caused by *Chlamydia pneumoniae* and *Mycoplasma pneumoniae*. Prophylactic penicillin (PCN) can reduce mortality related to pneumococcal infection in young children. Infants with SCD should be started on twice daily PCN and continued until they reach five years of age. Prophylactic PCN should be continued indefinitely in the event of splenectomy or invasive pneumococcal infection.[4,6] Patients with SCD should be vaccinated against *Streptococcus pneumoniae* and should also receive immunizations based on the Advisory Committee on Immunization Practices (ACIP) harmonized immunization schedule, unless personal contraindication dictate otherwise.[6]

Analgesics

Mild to moderate pain can often be treated at home with supportive care, such as rest, fluids, warm compresses, and OTC analgesics (e.g., NSAIDs, acetaminophen). [4,6] Management of severe pain associated with VOC requires timely administration of parenteral opioids by patient-controlled analgesia or frequently scheduled dosing. [6] Rapid pain management initiated within 30 minutes from triage for VOEs is a well-supported evidence-based guideline for treating SCD. Managing chronic pain should involve determining the cause and type of the SCD-related pain. The use of long- and short-acting opioids is recommended to manage chronic pain that is unresponsive to non-opioid analgesics. The patient's response to treatment, in terms of relief, adverse effects, and functional outcome should be evaluated to guide long-term use of opioids. [3]

Hydroxyurea

Hydroxyurea stimulates the production of fetal hemoglobin (HbF) nearly two-fold and decreases production of adult hemoglobin (HbA); this will then reduce the relative concentration of the sickle hemoglobin (HbS). Long-term use reduces the incidence of acute pain crisis, acute chest syndrome, and the need for blood transfusions. [4,6] It is indicated for adults with 3 or more moderate to severe pain crises in a one-year period, severe or recurrent acute chest syndrome, chronic symptomatic anemia, and sickle cell-associated pain that interferes with daily activities and quality of life. The use of hydroxyurea should be considered in all children older than 9 months of age regardless of disease severity. Refer to the 2014 Evidence-Based Report on the Management of Sickle Cell Disease for detailed recommendations on the use of hydroxyurea. [6]

Iron Chelation Treatment

Iron overload, which can lead to hemosiderosis, is a risk of chronic blood transfusions and often requires chelation therapy.[2] Though deferoxamine has been used in the past, newer oral chelating agents, deferasirox and deferiprone, are now more commonly used for outpatient purposes.[8]

New Drug Therapy

In July 2017, the FDA approved Endari™ (L-glutamine), the first treatment approved for patients with sickle cell disease in almost 20 years. The drug is for patients five years of age and older with SCD and is used to reduce severe complications associated with the disease.[9]

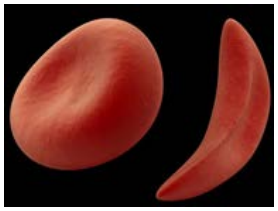


Table 1. Summary of SCD Complications:[2]

Acute Complications

- Infections
- Severe Anemia
- Vaso-occlusive crisis (VOC) or Vaso-occlusive episodes (VOEs)
  - Acute vaso-occlusive pain
  - Acute chest syndrome
  - Stroke
  - Priapism
  - Renal infarction
  - Dactylitis or Bone infarction
  - Myocardial infarction
  - Complications related to pregnancy
  - Venous Thromboembolism

Chronic Complications

- Chronic pain
- Avascular necrosis
- Pulmonary hypertension
- Renal impairment and hypertension
- Neurological deficits or seizure disorders
- Anemia
- Gallstones
- Chronic leg ulcers
- Delayed puberty and reduced growth
- Pregnancy complications and loss
- Proliferative retinopathy
- Osteoporosis and complications of bone infarction
- Cardiomyopathy with diastolic dysfunction
- Stuttering or recurrent priapism
- Hepatotoxicity (related to transfusional iron-overload or medications)
- Multi-organ failure (kidneys, liver, lungs)
- Morbidities associated with chronic pain (depression, anxiety, despair, insomnia, loneliness, helplessness, and dependence on pain medications)

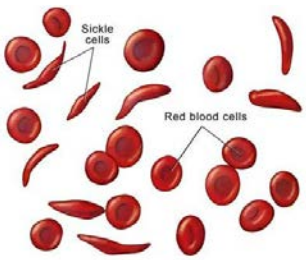


Table 2. Highlights of Evidence-based Recommendations for the Management of SCD[6]
(See full document for additional recommendations, further details regarding these highlights. and quality of evidence.)

Preventive services:

- Daily oral prophylactic penicillin up to the age of five years (strong)
- Pneumococcal vaccination for all ages (strong)
- Annual transcranial Doppler examinations from the ages two to 16 years in those with sickle cell anemia (strong)
- Referral to a specialist with expertise in long-term transfusion therapy to prevent stroke in children with conditional (170-199 cm/s) or elevated (≥200 cm/s) transcranial Doppler velocity (strong)

Addressing acute complications:

- Rapid initiation of opioids for treatment of severe pain associated with a vaso-occlusive crisis (strong)
- Use of incentive spirometry in patients hospitalized for a vaso-occlusive crisis (strong)

Addressing chronic complications:

- Use of analgesics and physical therapy for treatment of avascular necrosis (strong)
- Use of angiotensin-converting enzyme inhibitor therapy for microalbuminuria in adults with SCD (strong)

Caring for children and adults with proliferative sickle cell retinopathy:

- Referral to expert specialists for consideration of laser photocoagulation (strong)

Pulmonary Hypertension:

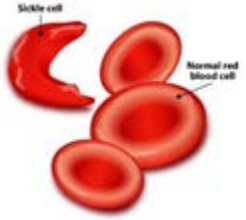
- Echocardiography to evaluate persons who have symptoms or signs suggestive of pulmonary hypertension (strong)

Hydroxyurea:

- Hydroxyurea therapy for adults with three or more severe vaso-occlusive crises during any 12-month period, SCD pain or chronic anemia interfering with daily activities, or severe or recurrent episodes of acute chest syndrome (strong)
- Optional treatment with hydroxyurea without regard to the presence of symptoms for infants, children, and adolescents (moderate)

Transfusion and Iron Overload:

- Preoperative transfusion therapy to increase hemoglobin levels to 10 g/dL in persons with sickle cell anemia (strong)
- Goal of maintaining sickle hemoglobin levels of less than 30% immediately prior to the next transfusion in children receiving long-term transfusion therapy (moderate)
- Assessment of iron overload (strong) accompanied by a recommendation to begin iron chelation therapy when indicated (moderate)



SCD Information Resources

CDC – Sickle Cell Disease
www.cdc.gov/ncbddd/sicklecell/index.html

American Sickle Cell Anemia Association
http://www.ascaa.org

NIH – What is Sickle Cell Disease?
https://www.nhlbi.nih.gov/health/health-topics/topics/sca/

Sickle Cell Disease Association of America
http://www.sicklecelldisease.org

Sickle Cell Information Center
http://scinfo.org

NIH - Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014
https://www.nhlbi.nih.gov/health-pro/guidelines/sickle-cell-disease-guidelines

NIH – Sickle Cell Disease Quick Guide
https://www.nhlbi.nih.gov/sites/www.nhlbi.nih.gov/files/Evd-Bsd_SickleCellDis_Rep2014.pdf

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3. Yawn BP, John-Sowah J. Management of sickle cell disease: recommendations from the 2014 expert panel report. *Am Fam Physician*. 2015;92(12):1069-1076.
4. Field JJ. Overview of the management and prognosis of sickle cell disease. In: UpToDate, Schrier SL (Ed), UpToDate, Waltham, MA. Accessed Oct 15, 2017.
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7. Hsieh MM, Fitzhugh CD, Weitzel RP, et al. Nonmyeloablative HLA-matched sibling allogeneic hematopoietic stem cell transplantation for severe sickle cell phenotype. *JAMA*. 2014;312(1):48-56.
8. Schrier SL, Bacon BR. Iron chelators: Choice of agent, dosing, and adverse effects. In: UpToDate, Menzer WC (Ed), UpToDate, Waltham, MA. Accessed Oct 15, 2017.
9. FDA approves new treatment for sickle cell disease. July 2017. https://www.fda.gov/newsevents/newsroom/pressannouncements/ucm566084.htm

ATTENTION PROVIDERS: PAYMENT ERROR RATE MEASUREMENT (PERM) FFY17 Currently Underway

Louisiana Medicaid is mandated to participate in the Centers for Medicare and Medicaid (CMS) **Payment Error Rate Measurement (PERM)** program which will assess our payment accuracy rate for the Medicaid and CHIP programs. If chosen in a random sample, your organization will soon receive a *Medical Records Request* from the CMS review contractor, CNI Advantage.

Please be advised that sampled providers who fail to cooperate with the CMS contractor by established deadlines may be subject to sanctioning by Louisiana Medicaid Program Integrity through the imposition of a payment recovery by means of a withholding of payment until the overpayment is satisfied, and/or a fine.

Please be reminded that providers who are no longer doing business with Louisiana Medicaid are obligated to retain recipient records for 5 years, under the terms of the Provider Enrollment Agreement.

For more information about PERM and your role as a provider, please visit the Provider link on the CMS PERM website: http://www.cms.gov/Research-Statistics-Data-and-Systems/Monitoring-Programs/PERM/Providers.html

If you have any questions, please call Catherine Altazan at 225-342-2612.

Online Medicaid Provider Manual Chapter Revisions as of October, 2017

Manual Chapter	Section(s)	Date of Revision(s)
Behavioral Health Services	Table of Contents Appendix F Wraparound Model 2.4 Coordinated System of Care 2.5 Record Keeping Appendix B Glossary and Acronyms	10/06/17 and 10/12/17 10/06/17 10/12/17 10/12/17 10/12/17
Durable Medical Equipment	18.4 Provider Requirements	10/25/17
Pharmacy	Table of contents 37.5 Covered Services, Limitations and Exclusions 37.11 Public Health Services 340B Program Appendix F Forms	10/13/17 10/13/17 10/13/17 10/13/17

Archived Online Medicaid Provider Manual Chapter Revisions as of October, 2017

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Remittance Advice Corner

Attention Fee for Service (FFS) Louisiana Medicaid Providers

Effective October 11, 2017, Fee-for-Service (FFS) Medicaid pharmacy claims will have Point of Sale (POS) edits for sumatriptan nasal powder (Onzetra®) and buprenorphine buccal film (Belbuca®). Please refer to www.lamedicaid.com for more information.

Attention Providers Of Immunization Services

An update to the fee for service claims processing system related to the processing and payment of immunization claims has been completed. Affected claims will be recycled for payment or denied for the correct edit without any action on behalf of the provider. The recycle will be included on the RA of Tuesday, October 31st, 2017.

For questions regarding this message and/or fee for service claims, please contact Molina Provider Relations at (800) 473-2783 or (225) 924-5040.

Updates to Healthy Louisiana related systems and claims processing changes are plan specific and are the responsibility of each health plan. For questions regarding Healthy Louisiana updates, please contact the appropriate health plan.

Attention Providers OF End Stage Renal Disease Services

Effective immediately, Louisiana Medicaid has updated the units of service on the fee for service procedure file for Healthcare Common Procedure Coding System (HCPC) code J1756-Injection, iron sucrose, 1 mg to align with the recommended dosage.

For questions regarding this message and/or fee for service claims, please contact Molina Provider Relations at (800) 473-2783 or (225) 924-5040.

Updates to Healthy Louisiana related systems and claims processing changes are plan specific and are the responsibility of each health plan. For questions regarding Healthy Louisiana updates, please contact the appropriate health plan.

For Information or Assistance, Call Us!

Provider Enrollment	(225)216-6370
Prior Authorization:	
Home Health/EPSDT – PCS	1-800-807-1320
Dental	1-866-263-6534 1-504-941-8206
DME & All Other	1-800-488-6334 (225) 928-5263
Hospital Pre-Certification	1-800-877-0666
Provider Relations	1-800-473-2783 (225) 924-5040
REVS Line	1-800-776-6323 (225) 216-(REVS)7387
Point of Sale Help Desk	1-800-648-0790 (225) 216-6381

General Medicaid Eligibility Hotline	1-888-342-6207
MMIS Claims Processing Resolution Unit	(225) 342-3855
MMIS/Recipient Retroactive Reimbursement	(225) 342-1739 1-866-640-3905
Medicare Savings Program and Medicaid Purchase Hotline	1-888-544-7996
For Hearing Impaired	1-877-544-9544
Pharmacy Hotline	1-800-437-9101
Medicaid Fraud Hotline	1-800-488-2917