SCD-Related Cardiopulmonary and Kidney Disease

- Evidence-based treatment guidelines for sickle cell disease complications that affect the heart, lungs, and kidneys.

W t atte

- SCD affects multiple organs that require patients to seek care from doctors that specialize in treating complications of the heart, lungs, and kidneys. It is important for these specialists to work closely with hematologists or SCD doctors to provide coordinated care for patients with SCD.

W t affect

H a d c a : Doctors will have access to guidelines that are based on the best available scientific evidence to improve their understanding of how to look for and treat complications of the heart, lungs, and kidneys, and make shared-decisions with patients.

SCD a : Their physicians will have more information and evidence-based guidance so that they can have informed discussions with patients to make the best testing and treatment decisions.

P c a : Policymakers will be informed of the knowledge gaps that exist in SCD evidence so they can direct research funding to fill in the gaps. In this regard, these guidelines may help shape policy.

W at a e t e g g t

H : The negative impact of hypertension on patient outcomes, particularly for African American individuals, means that a blood pressure goal of ≤130/80 mm Hg is the appropriate target to achieve in adults with sickle cell disease.

Sc : Patients with SCD who do not show any symptoms should not be routinely screened for pulmonary hypertension, abnormal lung function or sleep disorders. However, providers should carefully evaluate patients with SCD for signs and symptoms of cardiopulmonary disease that could suggest the need for diagnostic testing.

K d T a : Patients with SCD experiencing End-Stage Renal Disease or advanced chronic kidney disease requiring dialysis should not be excluded from consideration for kidney transplantation.

T ta be f a e ec e dat : 13

For more information on the 2019-2021 ASH Clinical Practice Guidelines on Sickle Cell Disease, t . e at g . g/SCDg de e .

SCD-Related Transfusion Support

- Clinical situations when transfusion should be used to care for people with sickle cell disease (SCD).

- People with SCD often require blood transfusions to prevent or treat organ damage associated with the disease. Additional clinical guidance may help physicians standardize and advance their patients' care and decrease side effects.

Pa ¬ ¬ SCD: To inform conversations with their physicians, particularly around when, where, how and why a transfusion should be provided.

B d be : To create an understanding of the more nuanced recommendations that affect how transfusions are administered.

H a : To provide a basis for guiding decisions for specific types of blood testing and circumstances for transfusion.

P c a : Policymakers will be informed of the knowledge gaps that exist in SCD evidence so they can direct research funding to fill in the gaps. In this regard, these guidelines may help shape policy.

- Patients with SCD who require transfusions should receive red blood cells that have undergone more extensive profiling that goes beyond traditional blood-type testing techniques.
- Therapies to suppress the immune system should be used under certain circumstances such as in patients with a sudden and pressing need for transfusion if they are at high risk of an immune response to the transfusion, a serious complication that can occur after a blood transfusion.
- The guidelines make additional recommendations on transfusion and SCD, including in:

T ta be f a e ec e dat : 12

For more information on the 2019-2021 ASH Clinical Practice Guidelines on Sickle Cell Disease, t . e at g . g/SCDg de e .

ASH guidelines are reviewed annually by expert work groups convened by ASH. Resources derived from guidelines that require updating are removed from the ASH website.

The American Society of Hematology (ASH) (*www.hematology.org*) is the world's largest professional society of hematologists dedicated to furthering the understanding, diagnosis, treatment, and prevention of disorders affecting the blood. For more than 60 years, the Society has led the development of hematology as a discipline by promoting research, patient care, education, training, and advocacy in hematology.

SCD-Related Cerebrovascular Disease

W at t c e

W t atte

W t affect

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SCD-Related Acute and Chronic Pain

- Evidence-based guidelines for the management of sickle cell disease (SCD)-related acute and chronic pain in children and adults.

W t atte

- Severe pain is the most common complication of SCD and affects individuals' quality of life. Acute pain episodes are the leading cause of emergency department visits and hospitalizations for individuals living with SCD. Further, chronic pain develops as individuals age and affects them daily.
- Acute and chronic pain management is a common clinical challenge for health care providers. This is in part due to the lack of strong evidence to support clinical decision-making.
- Health care providers may be unaware of all the available tools that can be used to manage acute and chronic SCD pain. These tools include both medications and treatments that are not medications.

W t affect

- **He at g t a d t e c ca d g a a age e t ca e:** Clinicians will have access to guidelines based on the best available scientific evidence to improve their understanding of how to look for, treat, and manage acute and chronic SCD pain.
- **E e ge c ca :** The guidelines make important recommendations for how quickly individuals living with SCD experiencing acute pain should receive medical attention.
- **P a Ca e a d Fa P c a :** Care of individuals living with SCD is often the responsibility of primary care and family physicians. This physician community has expressed a strong need for guidance and tools to help inform their care.
- **I d da g t SCD a d t e fa e be :** These guidelines provide an opportunity to make shared decisions with their health care providers regarding the management of their pain.
- **P c a e :** Policymakers will be informed of the knowledge gaps that exist in SCD evidence so they can direct research funding to fill in the gaps. In this regard, these guidelines may help shape policy.

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SCD-Related Acute and Chronic Pain

- **Ac te a :** Individuals seeking care for the treatment of acute pain should have their pain assessed and medication administered within one hour of their arrival at the acute care facility. Individuals should then be frequently reassessed every 30-60 minutes for consideration of additional doses of pain medication to optimize their pain control.
- **C c a :** Individuals who experience chronic pain may benefit from a tailored treatment plan when starting or ending chronic opioid therapy. Treatment decisions should balance the risks and benefits of opioids and consider the individual's function, goals, and durability of benefit over time.
- **C c a :** Medications that treat pain that are not opioids can be considered for individuals who experience chronic pain as part of a comprehensive pain isions

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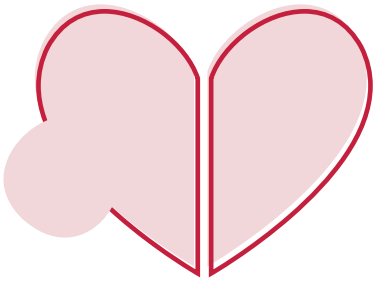

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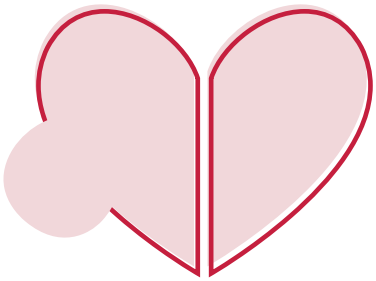

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