

COMMON CLINICAL DIAGNOSES AND CONDITIONS**SICKLE CELL DISEASE****INTRODUCTION**

Sickle cell disease is the most common autosomal recessive disease in African American individuals. It occurs in 1 in 625 live births to African-American couples. While it is most common in African Americans, it also occurs in individuals of Hispanic, Arabic, Native American and Caucasian heritage. Sickle cell disease results from a single base-pair substitution of thymine for adenine resulting in valine instead of glutamine in the sixth position of the Beta-globin molecule. Sickle cell disease results when this substitution occurs in a homozygous state. Less severe forms occur when the heterozygote state is combined with a second variant Beta-globin chain such as hemoglobin C or Beta^o-thalassemia. Clinical manifestations result from polymerization of the abnormal hemoglobin and “sickling” of the red cells. The clinical manifestations most important to pediatric hospitalists include recurrent and chronic pain from dactylitis and vaso-occlusive crises, acute chest syndrome, increased susceptibility to infections, aplastic crisis, splenic sequestration, cerebral vascular accidents and priapism. Pediatric hospitalists commonly encounter patients with known or suspected sickle cell disease and care for the various complications associated with the disease.

KNOWLEDGE

Pediatric hospitalists should be able to:

- Review the genetics and pathophysiology underlying the variants of sickle cell disease and their complications.
- Compare and contrast common sickle crisis presentations by age group.
- Explain the impact of newborn screening on preventative care.
- Describe the signs and symptoms of dactylitis, vaso-occlusive crisis, sepsis, acute chest syndrome, aplastic crisis, splenic sequestration, cerebrovascular accidents and priapism.
- Describe indications for hospital admission, and escalation to intensive care.
- Identify the goals of inpatient therapy, attending to both acute and chronic needs.
- Summarize the roles of members of a comprehensive clinical care team, such as patients, family/caregiver, subspecialty physicians, social worker, pharmacist, physical therapist, discharge planner, psychologist and others.
- Discuss the therapeutic options available for complications of sickle cell disease and describe the rationale for choosing a specific management plan.
- Explain the approach toward acute and chronic pain management.
- Cite reasons for transfer to a referral center in cases requiring pediatric-specific services not available at the local facility.

SKILLS

Pediatric hospitalists should be able to:

- Correctly diagnose sickle cell disease and/or its complications by efficiently performing an accurate history and physical examination, determining if key features of the disease are present.
- Order appropriate laboratory and radiographic testing based on history and physical examination findings.
- Create a comprehensive evaluation and management plan including the use of antimicrobial therapy, intravenous fluid hydration, pain management, transfusion therapy, and initiation of cardiovascular and pulmonary supportive care measures.
- Identify patients with worsening status and respond with appropriate actions.
- Consult subspecialists in a timely manner when appropriate.

ATTITUDES

Pediatric hospitalists should be able to:

- Communicate effectively with patients and the family/caregiver regarding the disease process, expectations of inpatient therapy and transition of care to the outpatient arena.
- Collaborate with subspecialists and the primary care provider and to ensure coordinated longitudinal care for children with sickle cell disease.

SYSTEMS ORGANIZATION AND IMPROVEMENT

In order to improve efficiency and quality within their organizations, pediatric hospitalists should:

- Collaborate with a multidisciplinary team consisting of subspecialty physicians, social workers, pharmacists, physical therapists, discharge planners and psychologists to improve quality of care, increase patient satisfaction and facilitate timely discharge from the acute care setting.
- Identify existing limitations for optimal care within the current hospital setting and work with hospital administration and community partners to develop and sustain appropriate referral systems and coordinated transfers of care.
- Lead, coordinate or participate in the development of coordinated discharge plans and programs in the local community.

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