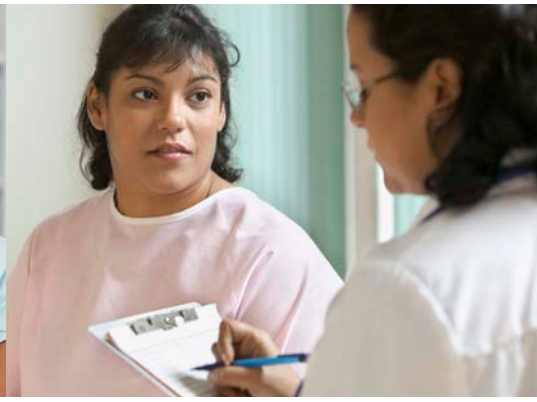
Evidence-Based Management of Sickle Cell Disease

Expert Panel Report, 2014: Guide to Recommendations





U.S. Department of Health and Human Services
National Institutes of Health
National Heart, Lung, and Blood Institute

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Acute Renal Failure (ARF)

1. In the setting of an acute rise in serum creatinine of ≥0.3 mg/dL,
   - Monitor renal function daily, including serum creatinine and fluid intake/output. **(Consensus–Panel Expertise)**
   - Avoid potential nephrotoxic drugs and imaging agents. **(Consensus–Panel Expertise)**
   - Evaluate the patient thoroughly for all potential etiologies in consultation with a nephrologist as needed. **(Consensus–Panel Expertise)**
2. Do not give blood transfusions to treat ARF unless there are other indications for transfusion. **(Consensus–Panel Expertise)**
3. Use renal replacement therapy (e.g., hemodialysis) when needed for acute renal failure. **(Consensus–Panel Expertise)**

Priapism

1. For an episode of priapism lasting 4 hours or longer, initiate interventions to include
   - vigorous oral or intravenous hydration and oral or intravenous analgesia **(Strong Recommendation, Low-Quality Evidence)***; and*
   - consultation with a urologist who can perform further evaluation and intervention for symptoms which do not remit with initial conservative medical management. **(Consensus–Panel Expertise)**
2. Do not use transfusion therapy for immediate treatment of priapism associated with SCD. **(Moderate Recommendation, Low-Quality Evidence)**

3. Consult with a hematologist for possible preoperative transfusion if surgical intervention is required.
(Consensus–Panel Expertise)

Hepatobiliary Complications

1. Treat acute cholecystitis in children and adults with SCD with antibiotics and surgical consultation.
(Consensus–Panel Expertise)

2. Treat asymptomatic gallstones with watchful waiting in children and adults with SCD. In those who develop symptoms specific to gallstones, treat with cholecystectomy. The laparoscopic approach is preferred if surgically feasible and available.
(Strong Recommendation, Moderate-Quality Evidence)

3. Consult with a hematologist or sickle cell expert for possible preoperative transfusion if surgical intervention is required.
(Consensus–Panel Expertise)

4. In children and adults with SCD and signs and symptoms of AHS or AIC, provide hydration, rest, close observation, and consult a sickle cell expert for further management.
(Consensus–Panel Expertise)

5. In children and adults with SCD and signs and symptoms of possible AHS or severe AIC, obtain urgent consultation with a sickle cell disease expert for diagnosis confirmation.
(Consensus–Panel Expertise)

6. In children and adults with SCD with confirmed AHS or severe AIC, perform simple or exchange transfusion.
(Consensus–Panel Expertise)