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PREGNANCY | Society for Maternal-Fetal Medicine

SMFM SPECIAL STATEMENT

Society for Maternal-Fetal Medicine Special Statement: Checklists for persons with sickle cell disease in pregnancy

Society for Maternal-Fetal Medicine (SMFM) | Nicolina Smith | Rebecca F. Hamm | Jessica Peterson | Alexandra DeRusso Forrest | Nicole A. Smith | Jourdan E. Triebwasser | SMFM Patient Safety and Quality Committee

Correspondence
The Society for Maternal-Fetal Medicine: Patient Safety and Quality Committee, PO Box 420016, Washington, DC 20042, USA.
Email: pubs@smfm.org

Abstract

This article presents checklists designed to guide clinicians in the comprehensive management of pregnant individuals with sickle cell disease, including prepregnancy, antepartum, delivery, and postpartum care, as well as the specific management of vaso-occlusive crises. The checklists aim to standardize care, enhance documentation and communication, and improve maternal and neonatal outcomes. Implementation strategies are discussed, emphasizing the need for integration of checklists into clinical workflows and continuous evaluation of quality indicators to address disparities and optimize patient care.

KEYWORDS

antenatal surveillance, disparities, fetal growth restriction, hemoglobinopathy, hydroxyurea, maternal morbidity, maternal mortality, pain management, perinatal outcomes, prepregnancy counseling, transfusion, vaso-occlusive crisis

1 | INTRODUCTION

Pregnant individuals with sickle cell disease (SCD) face increased risks of maternal and perinatal morbidity and mortality [1–6]. The checklists in this article aim to guide healthcare providers in managing pregnant patients with SCD in accordance with recommendations made by the Society for Maternal-Fetal Medicine (SMFM) and the World Health Organization, ensuring thorough risk assessment, vigilant monitoring, and collaborative care to optimize maternal and neonatal health [7, 8]. By following these checklists, healthcare teams can better navigate the complexities of SCD in pregnancy, with the ultimate goal of improving maternal and neonatal health outcomes.

Figure 1 contains a checklist for the management of SCD during the prepregnancy, antepartum, delivery, and postpartum periods. Figure 2 presents a checklist for the management of vaso-occlusive crises (VOCs). Practices should customize the checklists by adding or removing items as necessary to reflect local management protocols and further adapt them to meet individualized patient needs.

2 | SUGGESTIONS FOR IMPLEMENTATION

These checklists (Figures 1 and 2) are intended for use in both the inpatient and outpatient settings to increase the likelihood that obstetrical providers will perform all the

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First Encounter

An asterisk (*) denotes that if the patient presents for prepregnancy care, the task would be recommended at that time, but if not, then at the first prenatal encounter.

- ☐ * Evaluate health-related social needs.
- ☐ * Provide or refer for genetic counseling and assess partner status.
- ☐ * Review and update all vaccinations. If not previously documented and up to date, these vaccines are safe and recommended during pregnancy
  - o Pneumococcal vaccine
  - o Meningococcal vaccine
  - o Influenza vaccine
  - o COVID-19 vaccine
- ☐ * Recommend 4 mg/d folic acid in addition to prenatal vitamins without iron.
  - o Add iron only if deficiency is confirmed.
- ☐ Establish a multidisciplinary team that includes maternal-fetal medicine, hematology, genetics, pain management, behavioral health, and social work.
- ☐ * Establish plan for routine pain management as well as vaso-occlusive crisis.
- ☐ * Complete a detailed history and physical examination to assess for complications secondary to SCD.
- ☐ Assess baseline vital signs.
- ☐ Order baseline laboratory tests:
  - o CBC
  - o Iron studies
  - o Blood type and screen
  - o Complete metabolic panel
  - o Urine culture and urinalysis
  - o Urine protein
  - o Vitamin D level
- ☐ * Confirm dental examination within past year.
- ☐ * Confirm ophthalmologic examination within past year.
- ☐ Consider screening echocardiography if clinically indicated (see SMFM Consult Series #68). [7]
- ☐ Review medications for compatibility with pregnancy (eg, hydroxyurea, anticoagulants, iron chelators); (see SMFM Consult Series #68 Table 3). [7]
- ☐ Counsel on increased risk of preeclampsia.
- ☐ Begin low-dose aspirin at 12 to 16 weeks of gestation.

Antepartum Care

- ☐ Conduct urine assessment at each visit (urinalysis, dipstick, etc).
- ☐ Perform fetal growth surveillance every 4 weeks beginning at 28 weeks of gestation.
- ☐ If uncomplicated SCD and normal fetal growth, perform weekly or twice-weekly antenatal testing beginning at 32 weeks of gestation. (Individualize for those with complicated disease or pregnancy complications.)
- ☐ If alloimmunization is present, plan for blood product availability.

Delivery Planning

- ☐ Determine timing. (Consider delivery at 39 weeks or later unless otherwise indicated.)
- ☐ Determine mode of delivery. (Cesarean delivery should be reserved for obstetrical indications; avascular necrosis of the femoral head is not a contraindication to vaginal delivery.)
- ☐ Assess blood type and screen on admission; consider cross-match.
  - o If appropriate blood products cannot be obtained, consider transfer to a facility with blood product availability for delivery.
- ☐ Create labor and postpartum pain management plan, in collaboration with other specialties, such as anesthesia, hematology, and pain service.
- ☐ Seek neonatology consultation: address risk and management of neonatal opioid withdrawal.
- ☐ Develop a feeding plan, considering compatibility of restarting any prepregnancy disease-modifying medications. Breastfeeding can be encouraged.

Postpartum Care

- ☐ Consider prophylactic postpartum anticoagulation, particularly after cesarean delivery, or if other high-risk features are present. (eg, obesity, prior VTE).
- ☐ Coordinate transition of care (eg, hematology, any other supporting services).
- ☐ Provide contraceptive counseling. (LARC and progestin-only contraceptives are favored and should be offered.)

FIGURE 1 Sickle cell disease (SCD) in pregnancy checklist. CBC, complete blood count; COVID-19, coronavirus disease 2019; LARC, long-acting reversible contraception; VTE, venous thromboembolism.

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- ☐ Identify and minimize triggers (eg, infection, dehydration).
- ☐ Order laboratory/diagnostic tests:
  - CBC with differential
  - Lactate dehydrogenase
  - Comprehensive metabolic panel
  - Urinalysis
  - Urine culture
  - Chest X-ray if respiratory symptoms, hypoxia, or a significant decrease in Hgb
- ☐ Provide supportive care:
  - Hydration with isotonic fluids (PO or IV), while monitoring for signs of fluid overload
  - Supplemental O_2 as needed to maintain SpO_2 ≥95%
- ☐ Offer pain management:
  - Rapidly (within 1 hour of triage unit arrival) assess and administer analgesia; reassess frequently (every 30-60 minutes).
  - Tailor opioid dosing to the patient with consideration of their baseline opioid therapy and previous effective therapy.
  - Short courses of NSAIDs may be considered in the second trimester and postpartum.
  - Support nonpharmacologic therapies, including massage, yoga, transcutaneous electrical nerve stimulation, virtual reality, and guided audiovisual relaxation.
- ☐ Manage blood product use:
  - Perform RBC transfusion if Hgb is significantly lower than baseline or in cases of complicated VOC.
  - Conduct exchange transfusion in cases of acute ischemic stroke, ACS, hypoxic respiratory failure, or multisystem organ failure.
  - The goal of transfusion is HbS <30%.
- ☐ Provide VTE prophylaxis during hospitalization.
- ☐ Coordinate multidisciplinary care with intensivists, hematology, maternal-fetal medicine, and behavioral health providers.

FIGURE 2 Vaso-occlusive crisis (VOC) checklist. ACS, acute chest syndrome; CBC, complete blood count; HbS, hemoglobin S; Hgb, hemoglobin; IV, intravenous; NSAIDs, nonsteroidal anti-inflammatory drugs; PO, orally; RBC, red blood count; SpO_2, peripheral capillary oxygen saturation; VTE, venous thromboembolism.

unique tasks recommended for managing pregnancies complicated by SCD. Implementation of these checklists should start with the assembly of a motivated and qualified team. In this case, the team should include at least a physician (a maternal-fetal medicine subspecialist or a general obstetrician), an inpatient nurse, an outpatient nurse, and a practice manager. Institutions may also consider involving a hematologist, given the close collaboration with this specialty in the management of pregnancies affected by SCD. The team should consider how the checklist components can be implemented effectively across the outpatient and inpatient settings. General recommendations for implementing checklists can be found in the *Society for Maternal-Fetal Medicine Special Report: The Development and Implementation of Checklists in Obstetrics* [9].

3 | QUALITY INDICATORS FOR SCD IN PREGNANCY

Once the checklists have been implemented, it is essential to follow up to evaluate outcomes, identify areas for future improvement and checklist revisions, and consider optimizing clinical processes. Clinical quality indicators can be used to assess whether checklists are being used effectively. Some suggested quality indicators for pregnant individuals with SCD include the percentage of gravidas with SCD who had the following:

1. Completion of recommended vaccinations
2. Documentation of a pain management plan for VOC, including both pharmacologic and nonpharmacologic interventions
3. Recommendation for aspirin for preeclampsia prophylaxis
4. Regular fetal surveillance, including growth ultrasounds and antenatal testing, if indicated
5. Detailed delivery plan documented in the antepartum record, including timing and mode of delivery
6. Development of postpartum contraception plan

As the majority of patients with SCD identify as Black, optimal management of SCD is inherently an issue of health equity [10, 11]. Sites can consider evaluating the

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implementation of these checklists using quality improvement study designs as an intervention to reduce the impact of implicit biases and structural racism on care decisions around SCD. For guidance on health equity in quality improvement, refer to *SMFM Special Statement: How to Incorporate Health Equity Into Quality Improvement and Patient Safety Efforts* [12].

ORCID

Nicolina Smith https://orcid.org/0000-0003-1329-9956
Rebecca F. Hamm https://orcid.org/0000-0002-2587-2054
Jessica Peterson https://orcid.org/0000-0002-3708-1794
Alexandra DeRusso Forrest https://orcid.org/0000-0003-4573-1491
Jourdan E. Triebwasser https://orcid.org/0000-0002-1309-7935

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