UGANDA NATIONAL GUIDELINES FOR MANAGEMENT AND PREVENTION OF SICKLE CELL DISEASE

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Objectives of the Guidelines

- To guide health workers in standardized and simplified management of Sickle cell Disease (SCD) crises and complications at all levels of the health care system.

- To provide guidance on proper use of several prophylactic drugs among People Living with Sickle Cell Disease (PLSCD).

- To provide standardized and simplified guide on home basic care for PLSCD.

Objectives of the Guidelines Cont'd

- To provide a standardized and simplified guide on testing and confirmation of SCD and Newborn screening.
- To provide standardized and simplified guide on Genetic counseling of Sickle cell traits and PLSCD.
- To provide a guide on the management of pregnancy in sickle cell disease
- To act as a tool for training and creating awareness with regard to SCD.

Content:
The Guidelines have Seven Chapters

1. Clinical Feature of SCD
2. Diagnosis
3. Genetic Counseling
4. Management
5. Health Maintenance
6. **Pregnancy in Sickle Cell Disease**
7. Prevention

Chapter 6: Pregnancy in Sickle Cell Disease

- Complications of Pregnancy in SCD
- Preconception
- Prenatal screening and diagnosis
- Booking and Antenatal care
- Labour
- Caesarean section
- Postpartum care
- Contraception

Chapter 7: Prevention

- Community Involvement
  - Key community members
    - Spiritual Leaders
      - Cultural Leaders
      - Political Leaders
      - Village Health Teams

Chapter 7: Prevention

Screening –part 1

- Sample Collection

  - Materials for sample collection

  - Procedure for sample collection

  - Sample packaging and transportation

Screening – part 2

- Sample Processing

  - Sample reception

  - Data entry

  - Approval

  - Lab processing

  - Results printing and quality control

  - Results dissemination

- Premarital screening
- Pre-conceptual screening

These guidelines were presented before the MCH Cluster

The recommendations from the cluster are outlined below;

- Role of the Community in Sickle cell prevention and management
- Referral system in SCD management
- Creating in-depth guidelines in management of SCD other than these general guidelines

The Corrections done

Prevention

We added a full chapter on prevention and it contains;

- Role of the Community

- Roles of different stake holders
  - Cultural leaders
  - Spiritual leaders
  - Political leaders
  - VHTs

- Genetic counselling

- Pre-marital/Pre-conceptual screening

- Newborn screening

Referral Flow System

We added a section on the referral flow in management of SCD and carefully indicated what should be done at every level of healthcare.

- Health centre II's
- Health centre III's
- Health centre IV's
- District Hospitals
- Regional referral hospitals
- National Referral hospital

SCD IN PREGNANCY

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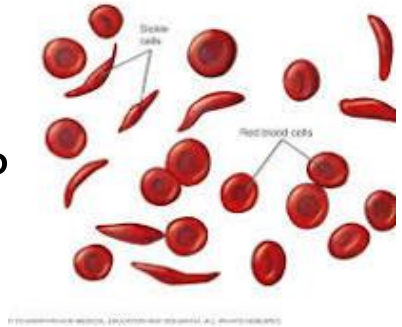
SCD IN PREGNANCY

- Introduction/ backgrc
- Preconception care
- ANC
- Intrapartum care
- Post natal care

SCD IN PREGNANCY

- Pathophysiology involves interplay of vaso-occlusion, hemolysis, inflammation, & altered blood flow
- increases risk of pregnancy complications e.g. SB, LBWT, P (Kassebum et al, 2017)

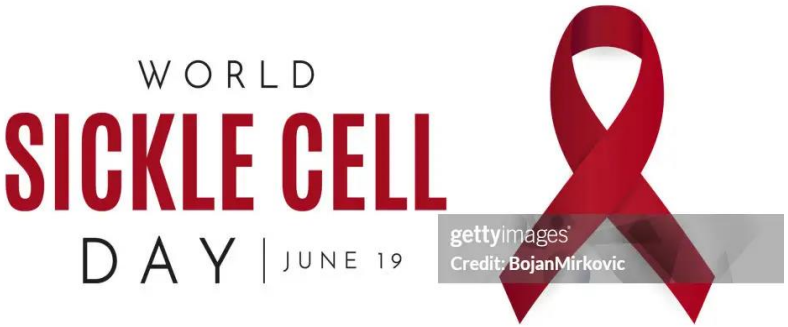


SCD IN PREGNANCY

- Prevalence in pregnant women still unknown
- Impact on and determinants of pregnancy outcomes not well documented in our setting
- No standardized guidelines on management in pregnancy to optimize outcomes
- Many interventions improve quality of life with many achieving reproductive potential
- Lancet Heamatolgy commission recommendations 1 &2 (epidemiological data 1 & 2, access to emerging treatments and interventions)
- Knowledge of determinants of outcomes improves risk stratification in the pregnant population
- Targeted interventions through development of protocols

PCC & Relevance

- contraception
- Medications
- Vaccinations
- Crisis avoidance
- Genetic counselling and couple screening
- Low threshold for seeking medical help



Information that is particularly relevant for women planning to conceive includes:

- The role of dehydration, cold, hypoxia, overexertion and stress in the frequency of sickle cell crises
- How nausea and vomiting in pregnancy can result in dehydration and the precipitation of crises
- The risk of worsening anaemia, the increased risk of crises and acute chest syndrome (ACS) and the risk of increased infection (especially urinary tract infection) during pregnancy.
- The increased risk of having a growth-restricted baby, which increases the likelihood of fetal distress, induction of labour and caesarean section
- The chance of their baby being affected by SCD
- An up-to-date assessment for chronic disease complications.

The assessment for chronic disease complications should include:

- Screening for pulmonary hypertension with echocardiography
- Blood pressure and urinalysis should be performed to identify women with hypertension and/or proteinuria.
- Retinal screening
- Screening for iron overload. In women who have been multiply transfused
- Screening for red cell antibodies

Medications & vaccinations during PCC

- Penicillin prophylaxis
- Vaccinations: Pneumococcal & Hep B
- **ACEIs** should be stopped pre conception
- Folic Acid supplementation
- **Hydroxy urea** should be stopped at least 3 months prior to conception, but if conceives on it, then counselling but do not terminate pregnancy.

Antenatal care

- Antenatal care by multidisciplinary team including an obstetrician and midwife with experience of high-risk antenatal care and a hematologist with an interest in SCD
- Aim to avoid precipitating factors of sickle cell crises such as exposure to extreme temperatures, dehydration and overexertion.
- Persistent vomiting can lead to dehydration and sickle cell crisis and women should be advised to seek medical advice early.

Appointment	Care for women with SCD during pregnancy
What should happen at at the first appointment?	Offer information, advice and support in relation to optimising general health (D)
Primary care or hospital appointment	Offer partner testing if not already done; review partner results if available and discuss PND if appropriate (D) Take a clinical history to establish extent of SCD and its complications Review medications and its complications; if taking hydroxycarbamide, ACE inhibitors or ARBs, these should be stopped (D) Women should already be taking 5 mg folic acid and antibiotic prophylaxis if no contraindication (D) Discuss vaccinations (D) Offer retinal and/or renal and/or cardiac assessments if these have not been performed in the previous year (D) Document baseline oxygen saturations and blood pressure Send MSU for culture
7–9 weeks	Confirm viability in view of the increased risk of miscarriage (D)
What should happen at the booking appointment?	Discuss information, education and advice about how SCD will affect pregnancy (D)
See midwife with experience in high-risk obstetrics if possible	Review partner results and discuss PND if appropriate (D) Baseline renal function test, urine protein/creatinine ratio, liver function test and ferritin should be

in high-risk obstetrics if possible	Baseline renal function test, urine protein/creatinine ratio, liver function test and ferritin should be performed (D) Extended red cell phenotype if not previously performed (D) Confirm that all actions from first visit are complete (D) Consider low-dose aspirin from 12 weeks of gestation (D)
16 weeks: see midwife plus multidisciplinary review	Routine as per NICE; repeat MSU Multidisciplinary review (consultant obstetrician and haematologist)
20 weeks : see midwife plus multidisciplinary team	Detailed ultrasound as per NICE antenatal guideline Repeat MSU Repeat FBC
24 weeks: see multidisciplinary team	Ultrasound monitoring of fetal growth and amniotic fluid volume. Repeat MSU
26 weeks: see midwife	Routine check including blood pressure and urinalysis
28 weeks: see multidisciplinary team	Ultrasound monitoring of fetal growth and amniotic fluid volume Repeat MSU Repeat FBC and group and antibody screen
30 weeks: see midwife and offer antenatal classes	Routine check including blood pressure and urinalysis
32 weeks: see multidisciplinary team	Routine check Ultrasound monitoring of fetal growth and amniotic fluid volume

34 weeks: see midwife	Routine check including blood pressure and urinalyis
36 weeks: see multidisciplinary team	Routine check Ultrasound monitoring of fetal growth and amniotic fluid volume Offer information and advice about: • timing, mode and management of the birth • analgesia and anaesthesia; arrange anaesthetic assessment • care of baby after birth
38 weeks: see midwife and obstetrician	Routine check Recommend induction of labour or caesarean section between 38 and 40 weeks of gestation
39 weeks: see midwife	Routine check and recommend delivery by 40 weeks of gestation
40 weeks: see obstetrician	Routine check and offer fetal monitoring if the woman declines delivery by 40 weeks of gestation

ACE = angiotensin-converting enzyme; ARB = angiotensin receptor blocker; FBC = full blood count (for the woman); MSU = midstream urine; NICE = National Institute for Health and Clinical Excellence; PND = prenatal diagnosis; SCD = sickle cell disease

Pain crisis in ANC

- **Avoid precipitants**
- **Fluids :** 60ml/Kg/24 hours, **risk of overload in preeclampsia**
- **Oxygen**
- **Analgesics,** Paracetamol for mild pain, NSAIDS only between 12 &28 weeks, morphine for severe pain .
- Avoid Pethidine , risk of seizures
- **Thromboprophylaxis** if admitted with low molecular weight heparine
- **Transfusion** : if HB less than 6gm/dl or fall of more than 2gm/dl from baseline (prophylaxis vs acute? , TAPS 2 TRIAL)

opiates, they should be nursed in an area where they can undergo hourly observations (Box 1).

Box 1. Outline of management of acute pain[64]

- Rapid clinical assessment
- If pain is severe and oral analgesia is not effective, give strong opioids (e.g. morphine)
- Give adjuvant non-opioid analgesia: paracetamol, NSAID (if 12–28 weeks of gestation)
- Prescribe laxatives, antipruritic and antiemetic if required
- Monitor pain, sedation, vital signs, respiratory rate and oxygen saturation every 20–30 minutes until pain is controlled and signs are stable, then monitor every 2 hours (hourly if receiving parenteral opiates)
- Give a rescue doses of analgesia if required
- If respiratory rate is less than 10/minute, omit maintenance analgesia; consider naloxone
- Consider reducing analgesia after 2–3 days and replacing injections with equivalent dose of oral analgesia
- Discharge the woman when pain is controlled and improving without analgesia or on acceptable doses of oral analgesia
- Arrange any necessary home care and outpatient follow-up appointment.

Blood should be cross-matched for delivery if there are atypical antibodies present (since this may delay the availability of blood), otherwise a 'group and save' will suffice. D

In women who have hip replacements (because of avascular necrosis) it is important to discuss suitable positions for delivery. ✓

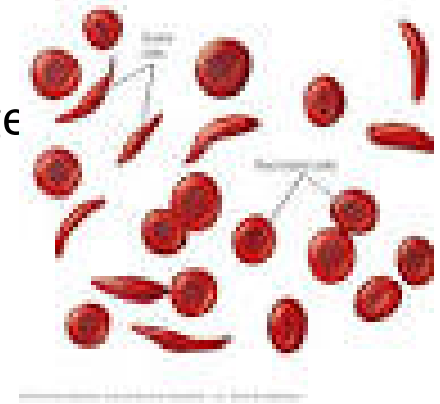
There are no randomised controlled trials to dictate the appropriate timing of delivery. Studies from the USA, UK, Jamaica and Africa have highlighted increased perinatal mortality, particularly during the later stages of pregnancy, in part owing to the complications of SCD.[21-26] The risks of abruption, pre-eclampsia, peripartum cardiomyopathy and acute sickle cell crisis are increased and unpredictable. It is the opinion of the developers that, like most 'high-risk' conditions, delivery of the baby at 38–40 weeks of gestation will prevent late pregnancy complications and associated adverse perinatal events.

Intrapartum care

- Timing of delivery (Between 38 and 40 weeks)
- Mode of delivery (vaginal unless has other obstetric indication for C/S)
- Place of delivery, hospital at bare minimum, high risk pregnancy.
- Multidisciplinary team
- Adequate warmth
- Fluids
- Pain management , Avoid Pethidine
- CTG for fetal monitoring
- SPO2 monitored, Oxygen supplement if less than 94%
- Regional anesthesia for C/S

Post partum

- New born screening if partner/ father was carrier
- Adequate Hydration with fluid balance chart until discharge
- Oxygen saturation monitored
- Pain mgt
- Low molecular weight heparin for 7 days post discharge if SVD and 6 weeks if C/S
- Contraception (progesterone only) , risk of DVT with estrogen containing pills



The risk of sickle cell crisis remains increased: in one study it occurred in 25% of women and was more common following general anaesthesia.[74] Hydration and oxygenation should be maintained and early mobilisation encouraged. Crises should be managed as for non-pregnant women. NSAIDs are routinely administered in the postpartum period and can be used during breastfeeding. Breastfeeding should be encouraged, as in women without SCD.

Thromboprophylaxis in the form of low-molecular-weight heparin is recommended while the pregnant woman is in hospital and for 7 days following vaginal delivery or for a period of 6 weeks following caesarean section.

Evidence level 4

7.2 What postpartum contraceptive advice should women be given?

This section should be read in conjunction with the Faculty of Sexual & Reproductive Healthcare guidance on postnatal hormonal contraception.[75] Contraceptive advice will often be the responsibility of primary care.

Progestogen-containing contraceptives such as the progesterone only pill (Cerazette®, Organon Laboratories Ltd, Haddesdon, UK), injectable contraceptives (Depo-Provera®, Pfizer Ltd, New York

B

SCD IN PREGNANCY

APWOYO MATEK

QSTNS, SUPPLEMENTS & CORRECTIONS

ARE WELCOME