Sultanate of Oman
Ministry of Health
The Royal Hospital
Department of Obstetrics and Gynecology

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Title: Sickle Cell Disease with Pregnancy

Introduction

Sickle cell disease (SCD) is a group of inherited single-gene autosomal recessive disorders caused by the 'sickle' gene, which affects hemoglobin structure.

The term SCD includes sickle cell anemia (HbSS) and the heterozygous conditions of hemoglobin S and other clinically abnormal hemoglobins. These include combination with hemoglobin C (giving HbSC), combination with beta thalassemia (giving HbSB thalassemia) and combination with hemoglobin D, E or O-Arab.

1.0 Pre pregnancy evaluation - Women with SCD are encouraged to seek consultation before their conception in the preconception clinic. They should be given information about the affects of the disease on pregnancy and how pregnancy affects SCD.

2.0 Evaluation at first visit

- Detailed history of the disease including complications, previous intensive care unit admission, previous hospitalizations and surgeries if any
- Course of previous pregnancies, outcome & any complications
- Husband's hemoglobinopathy status to be evaluated if not done
- Check if the woman is on Hydroxyurea, which increases the production of hemoglobin F. This should ideally be discontinued 3 months prior to pregnancy
- Iron chelators to be stopped at conception
- Assessment of cardiac function - Screening for pulmonary hypertension with echocardiography as the incidence of pulmonary hypertension is increased in patients with SCD
- Check for allergies to any medications
- Record blood pressure and check for proteinuria
- Booking ultrasound

2.1 Investigations at Booking

- Complete blood count
- Reticulocyte count
- Renal functions
- Liver functions
- Serum ferritin
- Antibody screening for the presence of any antibodies which may warrant the need for surveillance for fetal anemia
- Hb electrophoresis if not done in the past and baseline HbS levels
- Urine microscopy and culture

2.2 Advice / Counselling –

Provide women with information regarding the course of sickle cell disease in pregnancy-

- Role of dehydration, cold, hypoxia, overexertion and stress in increasing the frequency of sickle cell crises
- Dehydration possibly due to nausea and vomiting in pregnancy can lead to precipitation of crises
- Risk of fetal growth restriction. Sequestration in placenta may have risks to the fetus
- Genetic counselling regarding the baby being affected by SCD or trait depending on the sickling status of the partner.
- Possibility of increased risk of anemia and vaso occlusive crisis and worsening of the disease during pregnancy
- Increased risks of Chest infection and thromboembolism
- The patient should be advised to go to hospital and seek medical help early in case of any symptoms.

3.0 Antenatal care

- Patients with complications and recurrent crisis to be referred to the joint Hematology clinic for joint consultation with Hematologist

3.1 Obstetric Ultrasound:

- Dating scan at 11-14 weeks
- Anomaly scan at 18-20 weeks
- Growth scan at 28-30 weeks followed by 2-3 weekly till term.

3.2 Antenatal investigations

- Repeat CBC at each visit – result to be carefully followed. A marginally raised WBC count may be normal for these patients
- Urine analysis and culture is done at booking

- If initial culture is positive – treat and then screen monthly.
- If initial culture is negative, screen in each trimester.

3.3 Medications during pregnancy

- **Folic acid –** 5 mg daily through out pregnancy
- **Low dose Aspirin –** from 12 weeks onwards for the prevention of pre - eclampsia
- **Iron Supplementation-** To be given **only** if there is laboratory evidence of iron deficiency (which is unusual)
- **Penicillin Prophylaxis** - Penicillin V 250 mg BD in hyposplenic patients only
- **Prophylactic low-molecular-weight heparin** – To be started at 28 weeks gestation, Evaluate the need for thromboprophylaxis earlier in the presence of risk factors such as vomiting dehydration and immobilization and during hospital admissions prior to 28 weeks gestation. Postpartum thromboprophylaxis to be continued for all woman with sickle cell disease, duration depending on mode of delivery.
- **Non-steroidal anti-inflammatory** drugs - If required should be prescribed only between 12 and 28 weeks of gestation due to risk of premature closure of ductus arteriosus
- **Vaccines** - Polyvalent pneumococcal vaccine , meningo coccal vaccine , haemophilus influenza type B to be considered to be given for pregnant women

4.0 Timing and mode of delivery

- At 36 – 37 weeks of gestation, CBC, Hemoglobin S (HbS) to be checked and decision of blood transfusion or exchange transfusion to be considered prior to delivery to maintain HbS level <50 % and Hb more than 8gms/dl
- Spontaneous onset of labor can be awaited till term if there are no associated complications.
- Planned delivery at 38 weeks by induction of labor if indicated e.g. in cases of intrauterine growth restriction or recurrent crisis
- Vaginal delivery is preferred, and Caesarean section may be planned for obstetric indications
- Blood to be cross matched for delivery

4.1 Management in labor:

- Adequate hydration with IV fluids
- Epidural analgesia, Narcotic analgesics or Entonox inhalation
- A raised temperature (over 37.5ºC) requires investigation and to commence broad-spectrum antibiotics.
- Continuous fetal monitoring till delivery
- Prolonged labor to be avoided

4.2 In case of Cesarean delivery -

- Preoperatively, the patient should be well-hydrated
- If there is time, blood transfusion to achieve hemoglobin of 10 gms/dl & HbS level less than 40%.

- Exchange transfusion may need to be arranged in case of an elective Cesarean section. A pre anesthetic check should be done.

- Regional anesthesia is safer and preferable

5.0 Post partum care

- In pregnant women where the baby is at high risk of SCD (i.e. the partner is a carrier or affected), early testing for SCD should be offered
- Adequate hydration and analgesia until discharge
- Low-molecular-weight heparin while in hospital and till 10 days post-partum following vaginal delivery or for a period of 6 weeks following caesarean section
- The same level of care and vigilance should be maintained as has been described for antenatal care
- Contraceptive advice to be given -

  - Depot Medroxy progesterone acetate and Mirena are good options
  - COCs and IUCD are usually avoided
  - Barrier methods can be used but have high failure rate
  - Sterilization if she has completed her family

6.0 Management of crisis in pregnancy

6.1 Admission –

- Multidisciplinary team management involving Obstetricians, Hematologists and pain management team
- Examination to focus on the site of pain, any atypical features of the pain and any precipitating factors and signs of infection
- Investigations – CBC, RFT, LFT, HbS, urine culture to be done. Hemolytic markers may be sent if suspecting hemolysis

6.2 Hydration –

- Fluid of choice - 5% dextrose with 0.45% saline
- Required 3-4 L/day if cardiac status is normal.
- Fluid intake of at least 60ml/kg in 24 hours should be ensured

6.3 Pain management – Do a rapid clinical assessment

- **Mild to moderate pain** – Start with paracetamol. NSAIDs can be used between 12 and 28 weeks of gestation. Tramadol can be given.

- **Severe pain** – Patient may be referred to pain management team. Start parenteral narcotics e.g tramadol / morphine as per the pain management protocol
- Monitor pain, sedation, vital signs, respiratory rate and oxygen saturation until pain is controlled and signs are stable
- Prescribe laxatives, antipruritic and antiemetics if required
- Consider reducing analgesia after 2–3 days and replacing injections with equivalent dose of oral analgesia

6.4 **Protocol for parenteral morphine -**

- **Loading dose**- 0.1mgm/kg,
- After 15 minutes if no response, give a second dose of 0.05mgm/kg. Repeat in another 15 minutes

Maintenance infusion – rate of 0.04 mgm / kg / hr

(Syringe pump – 10 mgms (5 ampules) + 45 ml saline)

- **Extra boluses** of 0.01mg = 10 microgram/kg, can be given in case of break through pain
- Decrease infusion dose by 20% /day and stop when you reach <50% of recommended dose. Reduce the dose first before frequency.
- If pain increases while reducing the infusion rate, increase the oral analgesic dose. Oral analgesics should be given 4 - 6 hourly

6.5 Antibiotics - if evidence of infection. White blood cell counts are often raised in SCD and do not necessarily indicate infection

6.6 Thromboprophylaxis – If not already on heparin start thromboprophylaxis. All patients with SCD admitted to the hospital with an acute medical illness, including crisis, during pregnancy, should receive venous thromboembolism prophylaxis with low molecular weight heparin

7. Blood Transfusion-

- There is no absolute level at which transfusion should be undertaken and the decision must be made in conjunction with clinical findings
- Usually to treat acute complications or in preparation for surgery or delivery
- Aim to keep Hb more than 8gms and HbS between 30-50% at delivery
- Occasionally women may be on a prophylactic regular transfusion/exchange regimen, for the prevention of severe disease complications. This is decided by the Hematologist.

8. Indications for Exchange transfusion during pregnancy

- Acute chest syndrome and stroke
- Recurrent crisis or severe crisis with no response to 48 hours of conservative therapy with HbS level >60% and Hb >8 gms
- Hematologist opinion
- Hospital Policy for exchange transfusion to be followed

References

Title of book/ journal/ articles/ Website	Year of publication
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