
بسم الله الرحمن الرحيم

Management of sickle cell disease in pregnancy

Pre pregnancy evaluation

- Review clinical history of pain events and hospitalizations

evaluation generally includes:

- Confirmation of definitive diagnosis.

- Measurement of baseline blood pressure.

- Retinal evaluation to detect early proliferative sickle retinopathy

- Chemistry panel, LFT,RFT,, urinalysis, and ۲۴-hour protein

- the protein-to-creatinine ratio on a spot urine sample is also acceptable

●Hb / Hct and ferritin level. Full blood count , Hb electrophoresis , retic count , folate level,

● often have excessive iron stores, but a small proportion is iron deficient.

● Women with excessive iron stores → not receive iron ,consider delaying pregnancy ,use iron chelators

●Baseline urine culture, because of the increased asymptomatic bacteriuria and UTI

●Baseline pulmonary function tests, including pulse oximetry

- Hepatitis A,B and C , HIV, rubella screening
- Patient with multiple transfusion →cardiac magnetic resonance scaning , if iron deposition → delay pregnancy

- Echocardiogram as a screening test for pulmonary hypertension and early cardiac dysfunction

- Serologic red cell phenotyping (rh, kell, duffy, kidd and mns blood groups) and screening for red cell alloimmunization

  - to identify patients with multiple red cell alloantibodies who may be difficult to match for transfusion and may be at risk for hemolytic disease of the fetus and newborn .

  - If the woman has alloantibodies to red cell antigens, her partner should be tested for the corresponding antigens.

  - If the partner tests positive for the corresponding antigens, counseling concerning the risk of hemolytic disease of the newborn should be provided

- Testing partner for hemoglobinopathy

Management of medications and immunizations

- **Immunization** – Polyvalent pneumococcal, Haemophilus influenza type B, and meningococcal ,influenza vaccines are recommended for pregnant patients with SCD

- **Folic acid**
۵ mg/day has been recommended .

- **Hydroxyurea** – discontinue hydroxyurea three months before conception in both men and women

- Iron chelators – deferasirox and deferoxamine Both drugs are FDA pregnancy category C . Deferiprone is pregnancy category D.

- ACE inhibitors and ARBs – are teratogenic and should not be used during pregnancy.

●Prophylactic penicillin

- do not routinely initiate penicillin prophylaxis because of pregnancy but continue it in patients who are already taking prophylaxis.

- In England, penicillin prophylaxis is recommended based on the hyposplenism associated with SCD

- UK guideline→all adult with scd receive penicillin prophylaxy

Effect of sickle cell trait

- It is unclear whether SCT are at increased risk for pregnancy complications
- OR any modification to routine pregnancy care
- leg exercises, anti-embolism stockings, and compression devices for bed rest patient
- heparin use for high risk populations
- obtain a detailed history of any previous urinary abnormalities.
- If the history or initial urine screening tests are abnormal, perform frequent surveillance testing.
- It is important to test the patient's partner for hemoglobinopathy

MANAGEMENT DURING PREGNANCY

Prenatal care

- the level of care in pregnancy is the same for all type of these disorders

- monthly determination of hemoglobin level and chemistry panel.

- iron supplementation is avoided unless iron deficiency is documented by a low serum ferritin level

- Folic acid ۵ mg/day continued during pregnancy

- Control Nausea and vomiting of pregnancy , prevention of dehydration

- baseline and serial screening with urinalysis and culture

- For women with asymptomatic bacteriuria on initial urine culture, a course of antibiotic therapy and retest monthly until delivery.

- recurrent or persistent bacteriuria, suppressive therapy for the remainder of pregnancy.

- For women whose initial urine culture is negative, some clinicians screen each trimester, and others, rescreen monthly

●Close monitoring for development of preeclampsia

- low-dose aspirin from the beginning of the second trimester to ۵ to ۱۰ days before the expected date of delivery

● all hospitalizations should include VTE prophylaxis with an acute medical illness or obstetrical complication with LMWH or UFH

- VTE prophylaxis during the entire pregnancy in high-risk patients (such as those with a history of PE

- RCOG:LMWH from ۲۸ WK and post partum ۶wk.with other risk factor LMWH throughout pregnancy

	امتیاز
عوامل خطر مرتبط با شرایط طبی	
سابقه VTE قبلی (به جز موارد VTE به علت جراحی بزرگ)	٤
ترومبوفیلی اکتسابی (سندرم آنتی فسفولیپید آنتی بادی): حداقل یک معیار آزمایشگاهی و حداقل یک معیار بالینی	٤
سابقه VTE قبلی به علت جراحی بزرگ	٣
هر یک از مشکلات طبی: سرطان، بیماری قلبی، لوپوس فعال، پلی آرتروپاتی التهابی یا بیماری التهابی روده ، سندرم نفروتیک، دیابت ملیتوس نوع یک یا نفروپاتی، بیماری سیکل سل، اعتیاد تزریقی وریدی کنونی	٣
ترومبوفیلی ارثی پر خطر (کمبود آنتی ترومبین، کمبود پروتیین C یا S ، ترومبوفیلی کم خطر هموزیگوت یا همراه)	٣
ترومبوفیلی ارثی کم خطر (فاکتور ٥ لیدن هتروزیگوت، جهش ژن پروترومبین G٢٠٢١٠A)	١
تاریخچه خانوادگی VTE (بدون زمینه یا وابسته به استروژن) در بستگان درجه اول	١
وجود آنتی فسفولیپید آنتی بادی (فقط معیار آزمایشگاهی، بدون وجود معیار بالینی)	١
عوامل خطر مرتبط با شرایط عمومی	

●monitoring fetal growth with ultrasound and fetal well-being with non stress tests or biophysical profile in third trimester.

- A guideline from the United Kingdom suggests ultrasound examinations at weeks ۱۱ to ۱۳ to establish the estimated date of delivery and screen for Down syndrome

- weeks ۱۸ to ۲۰ to screen for congenital anomalies

- weeks ۲۸, ۳۲, and ۳۶ to screen for fetal growth restriction

- alloimmunization, should be assessed at the first prenatal visit.
- If negative initially, repeat at ۲۶ to ۲۸ weeks and at the time of delivery.
- the transfusion service should be notified so compatible blood is available for transfusion, if needed peripartum.
- The increased risk of alloimmunization in sickle cell disease does not change the standard recommendations for the use of anti-D immune globulin (eg, HyperRho S/D, RhoGAM) when appropriate

Management of acute painful episodes

- approach is similar to nonpregnant women
- except NSAIDs are generally avoided after ۳۰ weeks of gestation
- Opioids are the mainstay of treatment
- Oxygen supplementation , IV antibiotic ,hydration
- If IV AB not required →pnc prophylaxy should be ongoing
- Keep warm patient
- CBC,retic,biochemistry test,blood type,blood culture,ABG,PULSE OXIMETRY.
- Chest x ray if suspicious to chest syndrom

Prophylactic transfusion

- The use of prophylactic versus selective blood transfusions is controversial
- pregnancy outcome appear to be similar in both
- In patients with mild hemoglobin variants, or a benign clinical history, prophylactic transfusions are not utilized.
- prophylactic transfusion may be useful in SCD at highest risk of complications, (chronic renal, pulmonary, or hepatic disease), history of recurrent fetal loss, and in patients with either multigestational pregnancy or evidence of chronic fetal distress/intrauterine growth retardation

- such as those with previous perinatal mortality or severe anemia: Hb concentration <۶۰g/l and with acute anemia >۲g/dl falling of Hb

- anemia with cardiac or respiratory symptom
- on a chronic transfusion program
- in pregnant/postpartum women include prior to cesarean delivery and preeclampsia that does not improve after delivery

➢ transfusions on an outpatient basis every three to four weeks to keep hemoglobin ≥۹ g/dL and <۱۲.۰ g/dL and percent hemoglobin S below ۳۵ to ۴۰ percent with leukocyte-depleted matched for at least the C,D,, E, and Kell blood groups and CMV negative

Exchange transfusion is the recommended treatment for :

- acute chest syndrome
- stroke in SCD
- In chronically ill patients with high baseline hemoglobin levels, exchange transfusions are indicated to maintain the hemoglobin A level greater than ۳۰ to ۵۰ percent and the Hct greater than ۲۵%

Labor and vaginal delivery

- no medical contraindications to vaginal delivery
- UK:delivery beetwen ۳۸-۴۰ wk
- During labor and delivery the parturient should be kept well oxygenated (O_2 saturation ≥۹۵ percent), warm, and hydrated to prevent sickling.
- continuous fetal heart rate monitoring
- Neuraxial anesthesia is useful to reduce maternal cardiac demands secondary to labor pain and anxiety.
- Transfusion if HB<۸

Cesarean delivery

- If there is time, simple transfusion to achieve hemoglobin ۱۰ to ۱۱ g/dL in patients at increased risk of complications because of chronic lung disease, central nervous system disease, or multiorgan dysfunction.

- Fluid balance is important because risk for fluid retention from subclinical cardiomyopathy (۶۰cc Kg $24\ hr$

Postpartum VTE prophylaxis

- **Cesarean delivery**
  - Mechanical thromboprophylaxis
  - LMWH for a minimum of ١٠ days to selected low-risk individuals such as those with a non-HbSS genotype and no other risk factors for VTE.
  - LMWH for six weeks to patients with a HbSS genotype and/or a moderate to severe SCD history, older age, a history of pulmonary disease, an indwelling central venous catheter, a high platelet count, and/or a history of VTE or other VTE risk factors.
  - The RCOG recommends anticoagulation for all patients with SCD for six weeks following cesarean delivery

●Vaginal delivery

- pharmacologic thromboprophylaxis for five days after vaginal delivery, but continue in patients who remain hospitalized.

- The RCOG suggests LMWH prophylaxis for women in hospital and for seven days post-discharge

نتیجه ارزیابی	اقدام
مجموع امتیاز = ۲ یا بیشتر	تجویز داروی ضد انعقاد یا دوز پروفیلاکسی تا ۱۰ روز پس از زایمان توجه: در موارد سابقه VTE یا ابتلا به ترومبوفیلی ارثی یا اکتسابی با توجه به مدت یا مقدار تجویز دارو ممکن است بیشتر باشد باید با متخصص هماتولوژی یا داخلی نیز مشاوره شود
مجموع امتیاز = ۲	تجویز داروی ضد انعقاد یا دوز پروفیلاکسی حداقل تا ترخیص یا زمان تحرک کامل بیمار توجه: منظور از تحرک کامل یا Mobility این است که فرد در زمان بیداری، بیش از ۵۰ درصد اوقات در حال حرکت بوده و در بستر نباشد

- بیمار از ابتدا یا مدتي از بارداری داروی ضد انعقاد دریافت کرده تا ۶ هفته
- هپارین داد

- Following delivery, re-start hydroxyurea at the steady state dose the patient was on prenatally, if she is not breastfeeding.

- In women who were on iron chelators pre-pregnancy, iron status should be re-evaluated postpartum.

- all methods of combined (estrogen-progestin) and progestin-only hormonal contraception and the copper-releasing IUD safe and effective for women with SCD.


Thank you