Obstetric follow up in pregnant women with sickle cell disease

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Pregnancy in Women with Sickle Cell Disease

- ✓ Women with SCD are living longer with an improved quality of life
- ✓ They reach reproductive years and decide to have children
- ✓ Pregnancy is associated with an increased maternal and perinatal morbidity and mortality

Sickle Cell Disease

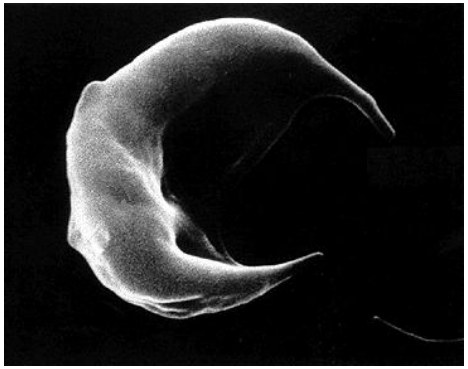
SCD is an autosomal recessive disorder caused by the 'sickle' gene, which affects haemoglobin structure

1. Sickle cell anemia (HbSS)
2. Heterozygous conditions :

   HbSC disease

   HbS-β thalassaemia

   HbD, HbE and HbO-Arab



SCD in Brazil

Prevalence

Sickle cell trait (HbAS)

General population: 4% (2 – 8%)

Afro descendents: 6 – 10%

200,000 children born each year

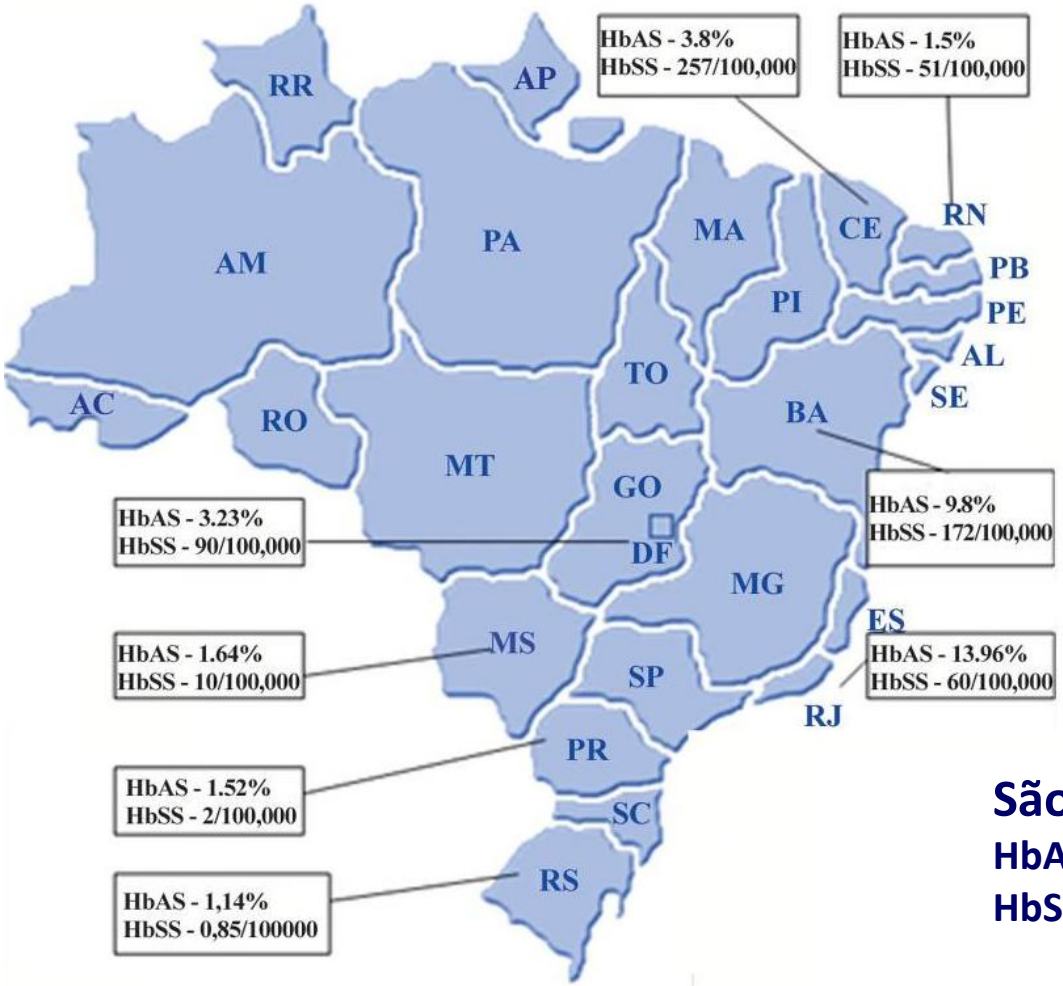
Estimative: 7,000,000 subjects

Sickle cell anemia (HbSS)

3,500 children born each year

Estimative: 25 – 30,000 subjects

Prevalence of sickle cell anemia and sickle cell trait in Brazil



São Paulo

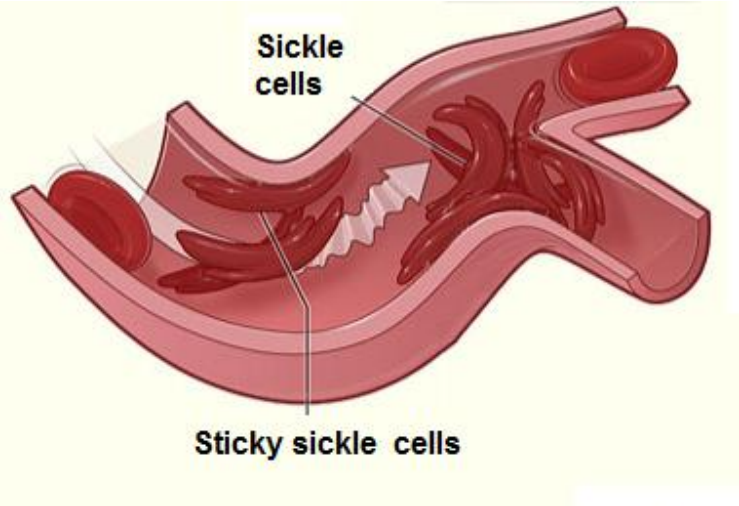
HbAS: 2,0 – 3,7%

HbSS: 4 – 15/100,000

Lervolino et al. Prevalence of sickle cell disease and sickle cell trait in national neonatal screening studies. Rev Bras Hematol Hemoter. 2011;33(1):49-54.

Sickle Cell Disease

Pathophysiology



- ✓ Abnormal Hb polymerases in low oxygen tension
- ✓ Haemolytic anemia
- ✓ Decreased red cell lifespan
- ✓ Increased adhesion of red cells to the endothelium
- ✓ Vaso-occlusion events: acute severe bony pain and multi-organ process
- ✓ Chronic multi-organ complications

Sickle Cell Disease

Clinical features

- Chronic anemia (Hb 6 to 9 g/dL)
- Mild genotypes may have higher Hb (HbSC)
- Recurrent episodes of severe bony pain (long bones, trunk)
- Variable frequency and severity of crises
- Increased risk of renal dysfunction, pulmonary hypertension, retinal disease, leg ulcers, cholelithiasis, stroke, and avascular necrosis

Sickle Cell Disease
Maternal morbidity and mortality

- ✓ Infections: pyelonephritis, pneumonia, septicemia
- ✓ Pulmonary hypertension
- ✓ Cholelithiasis
- ✓ Transfusion related complications: iron overload, infections (HIV, hepatitis B/C), alloimmunization
- ✓ Renal dysfunction
- ✓ Vaso-occlusion crises
- ✓ Stroke
- ✓ Retinopathy
- ✓ Avascular necrosis
- ✓ Chronic leg ulcerations
- ✓ Thromboembolic events
- ✓ Acute chest syndrome
- ✓ Pregnancy complications: pre-eclampsia/eclampsia , antepartum bleeding , placental abruption, cesarean, postpartum infections , blood transfusions
- ✓ Maternal death

Sickle Cell Disease

Maternal death

Author	Country	Description	N	Maternal deaths
Villers et al. 2008	USA	Retrospective database study 2000-03 multicenter (SCD)	17,952	0.05%
Alayed et al. 2013	Canada	Retrospective population-based 1999-2008 multicenter (SS 55%)	4,262	0.16%
Al Kahtani et al. 2012	Saudi Arabia	Retrospective (100% SS) 2001-10 Single institution	392	0.5%
Wilson et al. 2012	Ghana	Retrospective cohort 2007-08 Single institution (54% SS)	243	8.3%
Muganyizi & Kidanto, 2013	Tanzania	Local database study (SCD) 999-2001 Single institution	149	11.4%
Costa VMF 2012	UFMG Brazil	Prospective 2009-11 Single institution (50% SS)	60	3.3%
Nomura et al. 2010 + Paiva et al. not published	HCFMUSP Brazil	Retrospective 2001-08 + 2009-13 single institution (66% SS)	93	3.2%

Sickle Cell Disease Maternal complications

Author	Country	N	Preeclampsia	VTE	Cesarean	PPROM
Alayed et al. 2013 (SS 55%)	Canada	4,262	9.6%	1.2%	36.0%	3.2%
Al Kahtani et al. 2012 (SS 100%)	Saudi Arabia	392	9.7%	1.5%	16.1%	-
Wilson et al. 2012 (SS 54%)	Ghana	243	7.0%	-	48.8%	9.5%
Muganyizi & Kidanto, 2013 (SCD)	Tanzania	149	-	-	26.8%	-
Costa VMF 2012 (SS 50%)	UFMG Brazil	60	8.6%	6.7%	69.0%	-
HCFMUSP (SS 66%)	Brazil	93	16.1%	-	78.9%	-

Sickle Cell Disease Maternal complications

Author	Country	N	VO crisis	Pneumonia	Transfusion	Pyelonephritis	ACS
Al Kahtani et al. 2012 (SS 100%)	Saudi Arabia	392	43.1%	2.1%	37.2%	7.1%	1.0%
Wilson et al. 2012 (SS 54%)	Ghana	243	7.8%	0.8%	-	0.8%	-
Costa VMF 2012 (SS 50%)	UFMG Brazil	60	74.1%	-	58.6%	-	16.7%
HCFMUSP (SS 66%)	Brazil	93	46.2%	16.1%	33.3%	-	4.3%

Sickle Cell Disease

Perinatal morbidity and mortality

- ✓ Spontaneous miscarriage
- ✓ Fetal growth restriction
- ✓ Premature labor and delivery
- ✓ Low birth weight
- ✓ **Intrauterine fetal death**
- ✓ **Neonatal death**

Sickle Cell Disease
Intrauterine fetal death

Author	Country	Description	N	IUFD
Alayed et al. 2013	Canada	Retrospective population-based 1999-2008 multicenter (SS 55%)	4,262	1.2%
Al Kahtani et al. 2012	Saudi Arabia	Retrospective (100% SS) 2001-10 Single institution	392	4.8%
Wilson et al. 2012	Ghana	Retrospective cohort 2007-08 Single institution (54% SS)	253	11.5%
Muganyizi & Kidanto, 2013	Tanzania	Local database study (SCD) 999-2001 Single institution	149	25.7%
Costa VMF 2012	UFMG Brazil	Prospective 2009-11 Single institution (50% SS)	60	1.7%
Nomura et al. 2010 + Paiva et al. not published	HCFMUSP Brazil	Retrospective 2001-08 + 2009-13 single institution (66% SS)	90	10.0%

Sickle Cell Disease

Perinatal Results

Author	Country	N	Preterm <37w	BW<2500	SGA
Al Kahtani et al. 2012 (SS 100%)	Saudi Arabia	392	15.3%	18.6%	13.3%
Wilson et al. 2012 (SS 54%)	Ghana	243	23.3%	7.9%	-
Muganyizi & Kidanto, 2013 (SCD)	Tanzania	149	36.9%	34.2%	-
Costa VMF 2012 (SS 50%)	UFMG Brazil	60	44.8%	-	18.3%
HCFMUSP (SS 66%)	Brazil	93	28.6%	53.8%	30.0%

Sickle Cell Disease
Management of SCD in pregnancy

Preconception care

- Contraception
- Discussion about the complications of pregnancy in SCD
- Assessment for chronic disease, vaccination status
- Partner haemoglobinopathy status, genetic counselling
- Folic acid supplementation
- Medications (hydroxyurea), iron chelation therapy

Prenatal care

- Multidisciplinary care
- Screening for end organ damage, RBC antibody status (if it has not been undertaken before)
- Avoid precipitating factors of vaso-occlusion crises
- Early counselling to allow 1st trimester diagnosis (if the partner is a carrier)
- Vaccination and medications (folic acid, prophylactic antibiotics, aspirin, low molecular weight heparin)

Prenatal diagnosis for SCD - USA

- **Preimplantation genetic diagnosis (IVF)**
  to transfer an unaffected embryo or HLA identical to an affected sibling
  Kuliev et al. Preimplantationgenetic diagnosis for hemoglobinopathies. Hemoglobin. 2011;35:547-55
- **Noninvasive method: cell-free DNA from maternal plasma**
  Accurate sickle cell genotype: 75% (female) and 82% (male) fetuses
  Barrett et al. Digital PCR analysis of maternal plasma for noninvasive detection of sickle cell anemia. Clin Chem. 2012;58:1026-32
- **Invasive procedures**
  Chorionic villus sampling (CVS) 10-12 wks
  Amniocentesis after 15 wks
  ACOG. Practice Bulletin Nº 78: hemoglobinopathies in pregnancy. Obst Gynecol. 2007;109:229-37
- **75% of families with a HbSS fetus diagnosed prenatally chose to carry on the pregnancy**
  Alter BP. Prenatal diagnosis of hematologic diseases, 1986 update. Acta Haematol. 1987;78:137-41

Prenatal diagnosis for SCD - UK

Invasive procedures offered when both parents are carriers of unusual Hb

- ✓ Chorionic villus sampling 11th wks
- ✓ Amniocentesis 16th - 20th wks

Offer counselling about:

- understanding the test result
- making decisions about carrying on with the pregnancy or about termination in line with parent's beliefs and values

NHS Sickle Cell & Thalassaemia Screening Programme.
http://sct.screening.nhs.uk/

Prenatal routine

- ✓ Complete blood count
- ✓ Reticulocyte count
- ✓ Haemoglobin electrophoresis
- ✓ Antibody screening
- ✓ Ferritin
- ✓ Liver function
  (AST / ALT / BTF / DHL / GGT)
- ✓ Serology for hepatitis
  (hepatitis B and C, HIV, syphilis, toxoplasmosis, CMV, rubella)
- ✓ Renal function (U / C)
- ✓ 25-OH vitamin D
- ✓ Urinalysis / urine culture
- ✓ 24h proteinuria
- ✓ Cardiac function
  (echocardiography)
- ✓ Ultrasound
- ✓ Umbilical artery Doppler
- ✓ Retinal assessment
- ✓ Partner testing

Prenatal care - Medication

- ✓ **Folic acid** – daily (5mg/day)
- ✓ **Low-dose aspirin** (75mg/day from 12 wks): ↓ risk of preeclampsia
- ✓ **Hydroxyurea**
  - Teratogenic in animals
  - Congenital defects was not observed in limited human studies
  - Should be stopped during pregnancy and a level 3 ultrasound should be scheduled (termination is not indicated)
- ✓ **Prophylactic antibiotics** – penicillin therapy may be continued during pregnancy in patients who are already taking it
- ✓ **Iron supplementation** – only if there is laboratory evidence of iron deficiency (some patients are iron overloaded)
- ✓ **Low-molecular-weight heparin** – during antenatal hospital admissions

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Andemariam & Browning. Current management of sickle cell diasease in pregnancy. Clin Lab Med. 2013;33:293-310

Prenatal care – Nausea and vomiting

- ✓ Common in all pregnant women
- ✓ Can lead to dehydration and sickle cell crisis
- ✓ Women should be advised to seek medical advice early
- ✓ Prevention of dehydration may help to decrease the incidence of painful crises

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Hassell K. Pregnancy and sickle cell disease. Hematol Oncol Clin North Am. 2005;19(5):903-16

Prenatal care – urinary tract infection and asymptomatic bacteriuria

- ✓ Higher risk of urinary infections
  - Lower urinary tract: 40%
  - Upper urinary tract: 8% (pyelonephritis)
- ✓ Serial screening with urinalysis and culture (each trimester or monthly)
- ✓ Recurrent or persistent bacteriuria: antibiotic prophylaxis (Nitrofurantoin 100 mg/day)

Villers et al. Morbidity associated with sickle cell disease in pregnancy. Am J Obstet Gynecol. 2008;199:125 e1-e5.
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Prenatal care – preeclampsia

- ✓ Close monitoring: blood pressure and proteinuria should be assessed at each visit
- ✓ Low-dose aspirin (75mg/day from 12 wks): ↓ risk of preeclampsia

Duley et al. Antiplatelet agents for preventing pre-eclampsia and its complications. Cochrane Database Syst Rev. 2007;2:CD004659.

- ✓ Pre-existing proteinuria or known renal dysfunction: more frequent monitoring

Villers et al. Morbidity associated with sickle cell disease in pregnancy. Am J Obstet Gynecol. 2008;199:125 e1-e5.

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Prenatal care – alloimmunisation

- ✓ Screening for red cell alloimmunisation at the first prenatal visit
- ✓ If negative, some authors recommend to repeat at 24/28wks and at the time of delivery
- ✓ If positive: risk for haemolytic disease for the foetus and newborn
- ✓ Multiple red cell alloantibodies → difficult to match for transfusion

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Vichisnsky et al. Pregnancy in women with sickle cell disease. UpToDate. 2014. www.uptodate.com

Prenatal care – Fetal growth

- ✓ **Fetal growth monitoring with ultrasound**
- ✓ **Schedule of ultrasound scanning:**

  7-9w viability scan

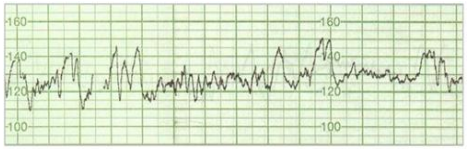
  11-14w routine 1st trimester scan

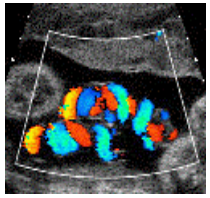
  20-24w anomaly scan

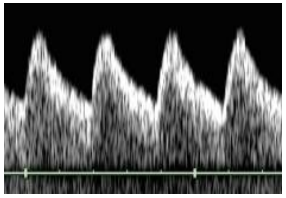
  every 4 weeks from 24w for serial fetal biometry
- ✓ **Umbilical artery Doppler Velocimetry**

  20-22w
- ✓ **Assessment of fetal well being**

  Cardiotocography and biophysical profile







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Prenatal care – Nutritional deficiencies

- ✓ **Children with SCA:** lower weight and height compared with reference standard and this deficit persists through adolescence and adulthood

- ✓ **Poor growth in SCA** results from a combination of relative hypophagia and increased metabolic demands

Reid. Nutrition and sickle cell disease. C R Biologies. 2013;336:159-63
Hyacinth et al. The role of nutrition in sickle cell disease. Nutr Metab Insights. 2010;3:57-67
Claster et al. Nutritional deficiencies in iron overloaded patients with hemoglobinopathies. Am J Hematol. 2009;84:344-8

- ✓ **Mothers with SCA** had lower weight and body mass index at first antenatal clinic and total weight gain was 6.9 kg for SS and 10.4 kg for AA controls

Thame et al. Pattern of pregnancy weight gain in homozygous sickle cell disease and effect on birth size. West Indian Med J. 2011;60:36-40.

Prenatal care – Nutritional deficiencies

Maternal undernutrition in SCD (HCFMUSP 2009-13)

- ✓ Pregestational BMI

  HbSS mothers → 14% underweight

  HbSC mothers → 5% underweight

- ✓ BMI at the end of pregnancy (Atalah et al. 1997)

  HbSS mothers → 50% underweight

  HbSC mothers → 16% underweight

Prenatal care – Blood transfusion

- ✓ **Routine prophylactic transfusion is not recommended during pregnancy**
  - Risks: alloimmunisation, delayed transfusion reactions, transmission of infection and iron overload
  - Prophylactic transfusion ↓ painful crises but did not influence fetal or maternal outcome
  - Systematic review: insufficient evidence to draw conclusions

RCOG. Management of sickle cell disease in pregnancy. Green-top guideline nº 61, July 2011
Okusanya & Oladapo. Prophylactic versus selective blood transfusion for SCD in pregnancy. Cochrane Database Syst Rev. 2013;12:CD010378

- ✓ **Blood should be matched for an extended phenotype** (C, D, E and Kell) **and leukocyte-depleted**

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Prenatal care – Blood transfusion

✓ **Prophylactic transfusion may be useful**

Women with previous perinatal mortality
Severe anemia (Hb<6g/dL)
Significant history of acute chest syndrome
Transfusion regimen before pregnancy for stroke prevention
Twin pregnancies

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Prenatal care – Blood transfusion

- ✓ **Transfusion for acute complications**
  - Acute chest syndrome (exchange transfusion)
  - Acute stroke (exchange transfusion)
  - Acute multi-organ failure
  - Acute anemia: Hb< 6g/dL or ↓ 2g/dL from baseline (increased haemolysis, splenic sequestration, red cell aplasia)
  - Reticulocytopenia (Parvovirus B19 infection or any infecction)
  - Severe, refractory pain crisis (> 10 days)
  - Prior to cesarean

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Parrish & Morrison. Sickle cell crisis and pregnancy. Semin Perinatol. 2013;37:274-9

Management of acute painful crisis

- ✓ **Increased incidence** during pregnancy: 27 – 50%
- ✓ **Multidisciplinary** team
- ✓ **Rapid clinical assessment for complications:** ACS, sepsis or dehydration
- ✓ **Precipitants factors:** infection, cold environment, excessive exercise, dehydration, hypoxia and stress
- ✓ **Fluids and oxygen** administered if required
- ✓ Initial **analgesia** should be given within 30 min
- ✓ Effective **analgesia** should be achieved within 1 h
- ✓ **Tromboprophylaxis** should be given

RCOG. Management of sickle cell disease in pregnancy. Green-top guideline nº 61, July 2011

Management of acute painful crisis

- ✓ **Examination should focus:** site of pain, any atypical features, precipitating factors, signs of infection, oxygen saturation

- ✓ **Initial investigations:** CBC, reticulocyte count, renal function, urine culture, blood culture, chest X-ray, liver function tests

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Management of acute painful crisis

Analgesia

- ✓ **Mild/moderate pain**

  Weak opioids (paracetamol + codein/ tramadol)
  NSAIDs (between 12 and 28w)

- ✓ **Severe pain**

  Morphine intermittent IV bolus 1-2 mg or patient-controlled administration system
  Pethidine should be avoided: toxicity and seizures

- ✓ **Monitor:** pain, sedation, vital signs respiratory rate and oxygen saturation every 20-30 min intervals

- ✓ ↓ **analgesia** after 2-3d and replace with equivalent oral dose

RCOG. Management of sickle cell disease in pregnancy. Green-top guideline nº 61, July 2011

Management of acute painful crisis

Fluids

- ✓ Oral or IV:
  - 60 mL/kg/h
  - 0.5 – 1.0 L of normal saline over 1st h and then 125mL/h (in the absence of cardiopulmonary disease)
- ✓ Careful fluid balance

Oxygen

- ✓ Oxygen saturations should be monitored
- ✓ Facial oxygen 3-6 L/min if < 95% or >3% below baseline

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Management of acute painful crisis

Antibiotics

- ✓ Infection is present in up to one-third of patients with vasoocclusive crisis
- ✓ If suspected: broad spetrum antibiotic thepapy after cultures are obtained:

  Initial choice → ceftriaxone (for encapsulated organisms: Streptococcus, Meningococcus and Haemophilus)

  Meningitis → vancomycin

Therapeutic blood transfusion

- ✓ Symptomatic patients who are unresponsive to conservative management
  (relapse < 7d, no improvement > 10d)

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Management of acute painful crisis

Assessment of fetal well being

- ✓ Cardiotocography
- ✓ Biophysical profile score
- ✓ Amniotic fluid volume

ACOG. Practice Bulletin Nº 78: hemoglobinopathies in pregnancy. Obstet Gynecol. 2007;109:229-37

- ✓ Corticosteroids should be considered in patients at viable GA and increased risk of preterm labor

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Acute Chest Syndrome

- ✓ **Incidence**: 7 to 20% of pregnant women with SCD
- ✓ **Clinical features:**
  fever, tachypnoea, chest pain, cough, shortness of breath, hypoxia, in the presence of new infiltrate on X-ray (~ H1N1 virus)
- ✓ **Pulmonary vascular occlusion** and **bone marrow emboli**
- ✓ **Maternal mortaty** rate of around 50%
- ✓ **Treatment**: IV antibiotics, oxygen, blood transfusion or exchange transfusion, respiratory support, critical care unit
- ✓ **Low-molecular –weight heparin** when pulmonary embolism is suspected

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Acute stroke

- ✓ **Infarctive or haemorrhagic**
- ✓ **Clinical features:**
  acute neurological impairment
- ✓ **Urgent brain imaging should be performed**
- ✓ **Emergency exchange transfusion**
- ✓ There is no evidence of benefit with thrombolysis

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Acute anemia

- ✓ **Splenic or hepatic sequestration**
- ✓ **Cholecystitis**
- ✓ **Parvovirus b19 infection**

- ✓ **Tratment:**

Blood transfusion

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Intrapartum care

- ✓ **Timing and mode of delivery**: 38+0w induction of labour
- ✓ **Cesarean only for obstetric indications**
- ✓ **Blood cross-matched for delivery**
- ✓ **Discuss positions for delivery in women who have hip replacements**
- ✓ **Risks of abruption, pre-eclampsia, peripartum cardiomyopathy and vasooclusive crisis are increased and unpredictable**

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Optimal care during delivery

- ✓ Give birth in units able to manage the complications
- ✓ Keep the woman warm and well hydrated (oral or IV)
- ✓ Pulse oximetry should be used
- ✓ Supplemental oxygen therapy (Sat>94%)
- ✓ There is no evidence for the use of prophylactic antibiotics
- ✓ Continuous FHR monitoring
- ✓ Routine thromboprophylaxis
- ✓ Epidural analgesia

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Postpartum care

- ✓ **Acute sickle pain is common (7 – 25%)**
- ✓ **Monitoring of fluid balance and oxygenation (sat > 94%)**
- ✓ **Early mobilisation and NSAIDs**
- ✓ **Close maternal surveillance for infection**
- ✓ **Thromboprophylaxis** while in hospital and 7d post-discharge following vaginal delivery and 6 weeks following cesarean
- ✓ **Breast feeding shoul be encouraged**
- ✓ **Early testing for SCD in newborn**
- ✓ **Postpartum contraceptive: progesterone only pill, injectable, and levonorgestrel intrauterine system**

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Ministério da Saúde, Brasil

Promote a change in the natural history of sickle cell anemia in Brazil, reducing the rate of mortality and sickness of these people.

Obrigada!