

Perioperative Management and Urgent Cesarean Delivery of a Complicated Patient with Severe Sickle Cell Disease and Acute Fatty Liver

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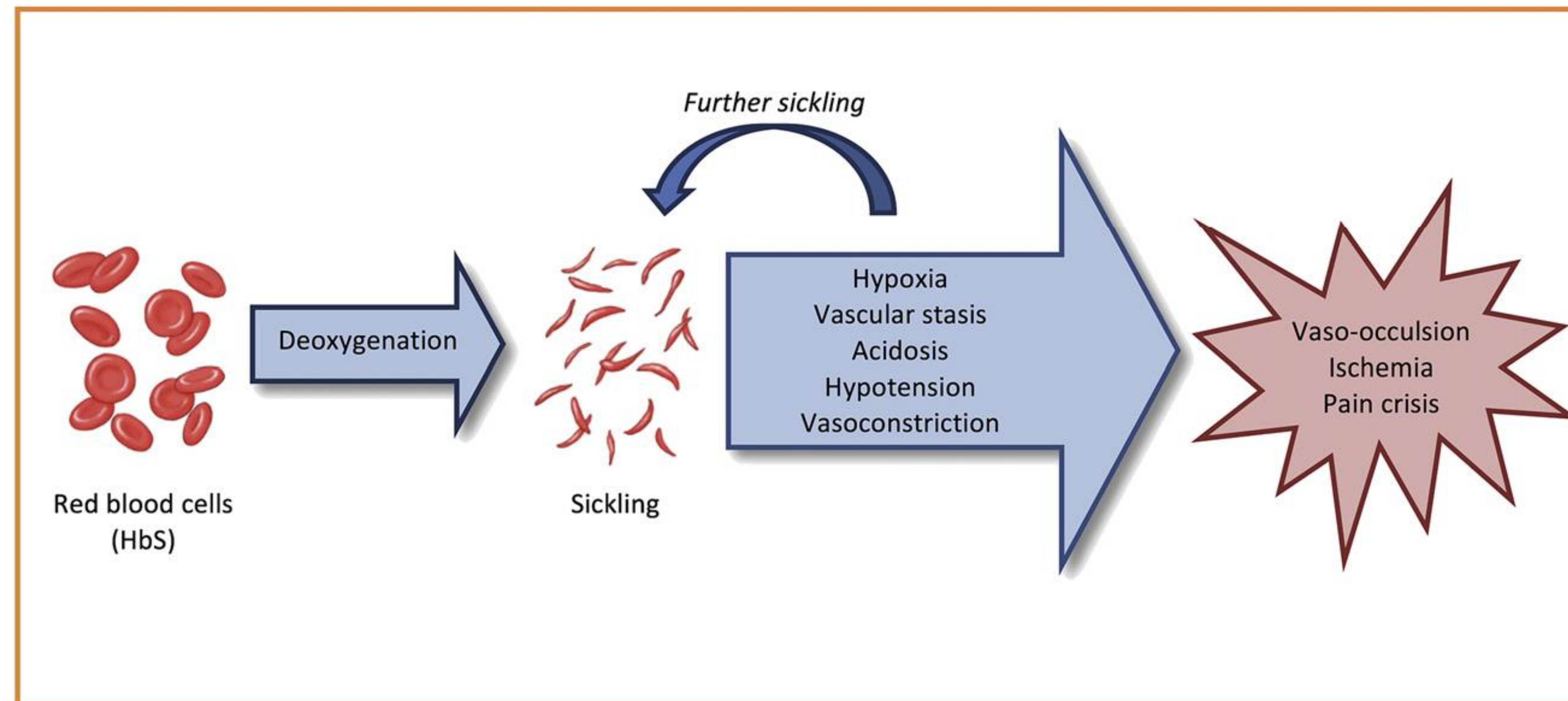
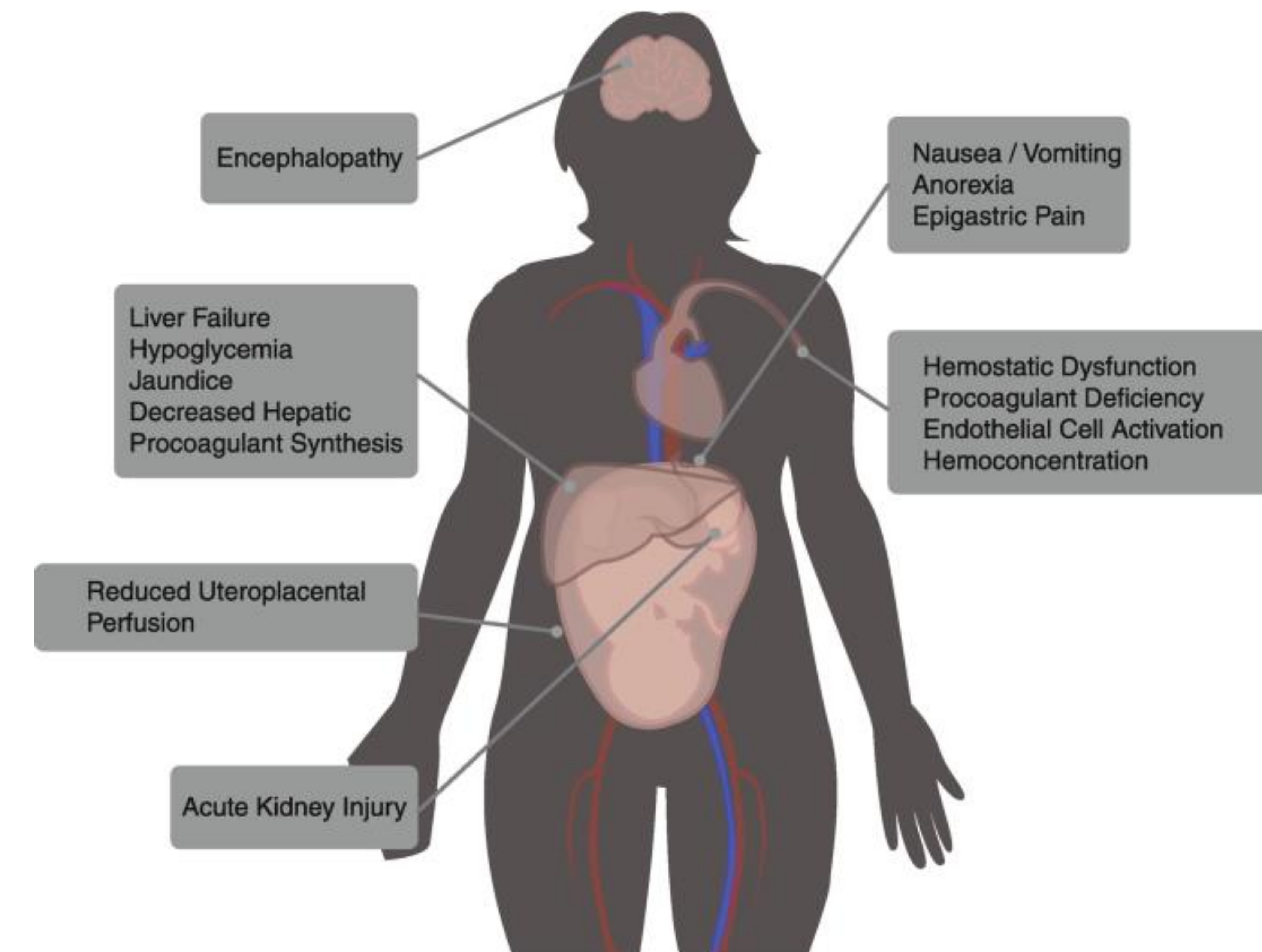


FIGURE 1. Sickling progression to pain and ischemia. HbS, hemoglobin S.

Stanley and Christian. *Sickle Cell Disease. J Oral Maxillofac Surg* 2013.



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INTRODUCTION

- Sickle cell anemia is caused by a homozygous mutation of the hemoglobin beta gene on chromosome 11p14.5 as an evolutionary response to malaria (1).
- Those with SCD present with chronic anemia and painful ischemic vaso-occlusive crises because these abnormal red cells sickle in an oxygen depleted environment result in impaired microcirculation and bone marrow ischemia (2).
- Pregnancy complicated by SCD is associated with increased risk of preeclampsia, eclampsia, gestational diabetes, chronic fetal hypoxia, IUGR, and spontaneous abortion(3).
- The normal physiological changes of pregnancy in SCD may result in an increased incidence of vaso-occlusive crisis, acute chest syndrome, osteonecrosis, hepatic necrosis, leg ulcers, thromboembolic events, chronic fetal hypoxia, IUGR, and spontaneous abortion (4).
- We present the peripartum management of a patient with severe SCD and multiple comorbidities who required cesarean delivery for worsening SCD, development of acute fatty liver of pregnancy, and non-reassuring FHR trace at 31 weeks gestation.

PATIENT INFORMATION

- 35 year old G1P0101, 4’ tall, 66.2 kg, BMI 44.25, ASA PS IV
- Past medical history: severe sickle cell disease, pulmonary hypertension (on home O₂), chronic osteomyelitis of b/l lower extremities.
- Abdominal compartment syndrome post cholecystectomy/splenectomy with chronic adhesions
- Multiple episodes of DVT with pulmonary emboli and chronic pyelonephritis
- Home medications: suppressive antibiotic therapy (ciprofloxacin) for chronic osteomyelitis and pyelonephritis, apixaban for DVT, hydroxyurea for SCD, and hydromorphone for chronic pain

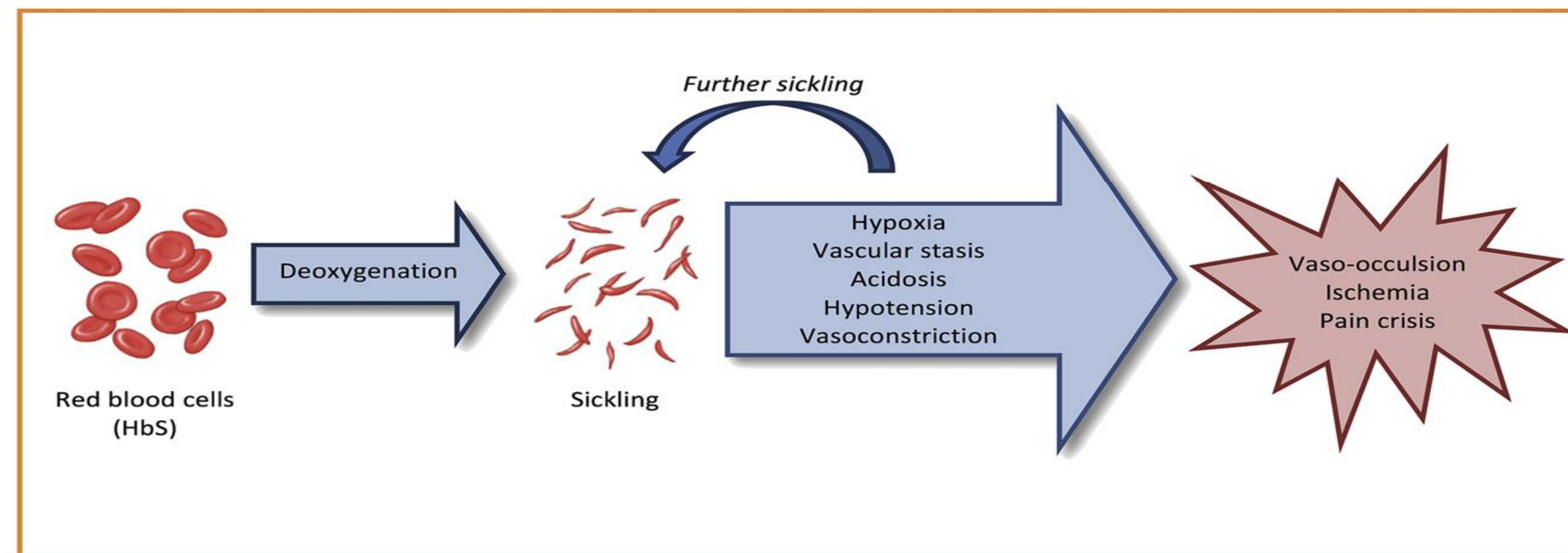


FIGURE 1. Sickling progression to pain and ischemia. HbS, hemoglobin S.

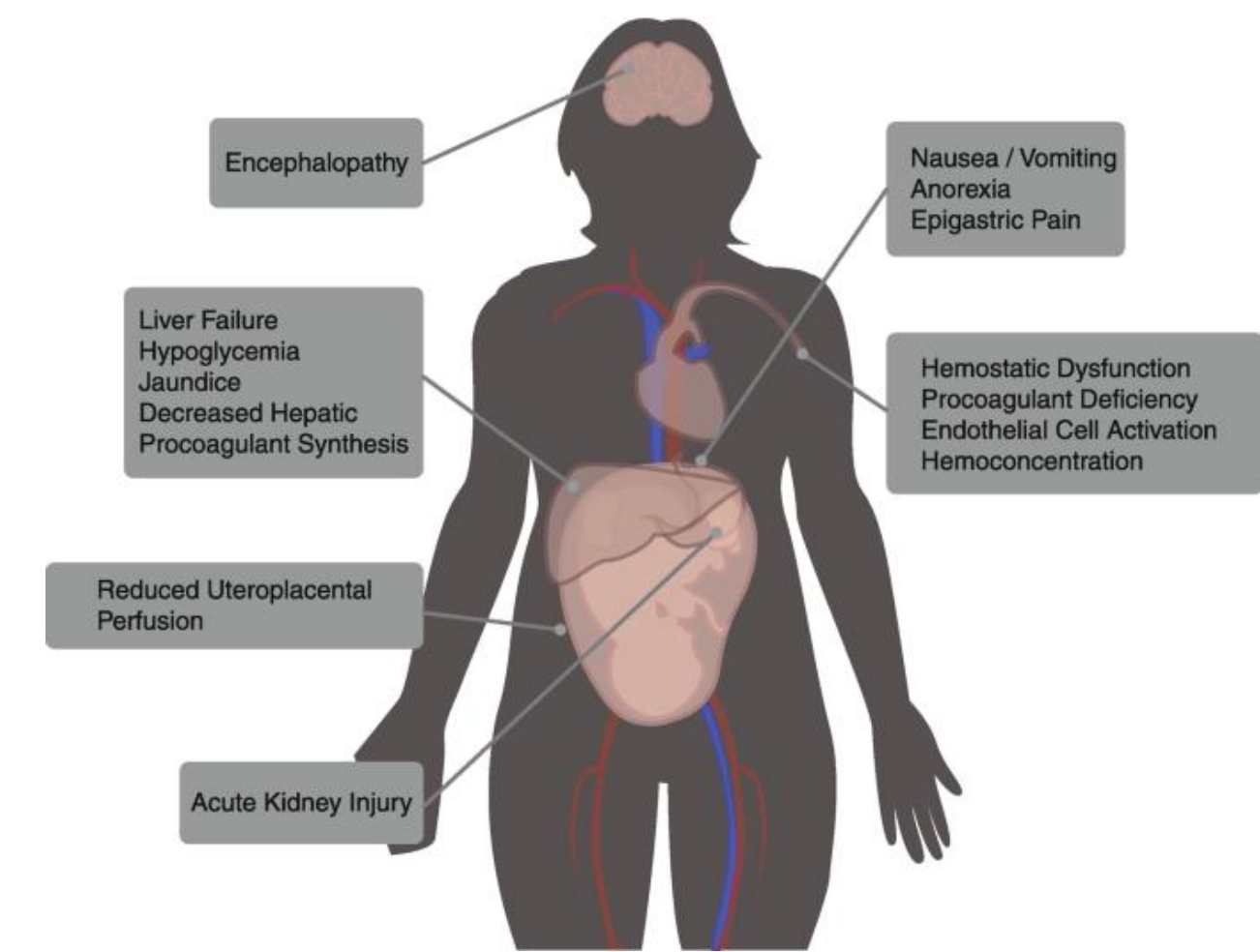
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CASE DESCRIPTION

- Presented with sickle cell crisis, found to be pregnant at 14 weeks 3 days gestation
- Ciprofloxacin (category C) and hydroxyurea (Category D) discontinued
- Diagnosed with right saphenous DVT and bilateral soft tissue skin infections in lower extremities from chronic osteomyelitis requiring bilateral above knee amputations
- Bilateral femoral and sciatic nerve blocks placed prior to general anesthesia for amputation procedures, ketamine (category N, not classified) used intra- and postoperatively to help control perioperative pain
- 19 weeks 6 days gestation: discharged to an inpatient rehabilitation facility
- 25 weeks 2 days gestation: developed altered mental status, fetal tachycardia, readmitted with urinary retention from Klebsiella urinary tract infection, and Covid 19 pneumonia, treated with nitrofurantoin (category B)
- Maternal ECHO showed severe pulmonary hypertension most likely from SCD and chronic DVTs/PEs
- Right heart catheterization deferred due to acute sepsis, discharged to home with physical and occupational therapy, on antibiotic regimen
- 29 weeks 4 days gestation: readmitted for acute sickle cell crisis and UTI, placed on IV antibiotics, IV hydromorphone
- 31 weeks gestation: developed acute fatty liver of pregnancy, creatinine increased from 0.55 mg/dL to 1.37 mg/dL, LFTs increased, became oliguric, non reassuring FHR trace
- URGENT CESAREAN DELIVERY!!!
- Multidisciplinary team consisting of Obstetrics, MFM, cardiology, infectious disease, and anesthesiology formulated care plan
 - Poor IV access, 9.5 F introducer, arterial catheter were placed
 - Hgb 7.3 mg/dL, Two units of PRBCs given
- Anesthetic plan
 - GA, transversus abdominis plane (TAP) blocks (INR 1.7)
 - Propofol, lidocaine, rocuronium (K 5.1), fentanyl, RSI
 - Intubation with glidescope blade #3.
 - Required phenylephrine infusion
 - Sevoflourane until the fetus delivered (APGAR 5¹,9⁵) followed by propofol infusion
 - No nitrous oxide (severe pulmonary hypertension)
 - Required 1 ampule of Sodium bicarbonate (50 mEq) for pH 7.1
 - Ketamine infusion (10 mg/hr) intra and postoperatively
 - TAP blocks attempted, distorted abdominal anatomy precluded successful placement
- Discharged with home care 8 days later.

DISCUSSION

- Sickle cell disease began as an evolutionary response to malaria and is the most common inherited hemoglobinopathy (1).
- The normal physiological changes of pregnancy in SCD overburden organs that have already sustained chronic injury, resulting in increased incidence of vaso-occlusive crisis, acute chest syndrome, osteonecrosis, hepatic necrosis, leg ulcers, thromboembolic events, chronic fetal hypoxia, IUGR, and spontaneous abortion (4).
- Hydroxyurea, a myelosuppressive agent, reduces Hgb SS and painful crises but is teratogenic. Our patient had worsening of her SCD after it was discontinued.
- Infection is more common with SCD. Osteomyelitis in our pt’s LE resulted in bilateral AKAs and potential phantom limb pain (central sensitization and peripheral hypersensitivity of cut nerve endings). Perineural infusion of local anesthetics are effective so we performed single shot femoral/sciatic blocks (5).
- Patients with SCD may develop precapillary pulmonary hypertension resulting from vasculopathy and a nitric oxide deficiency state (6). Labor also increases pulmonary vascular resistance. Unfortunately, there are no medications that lower pulmonary pressure in SCD patients with precapillary pulmonary hypertension. Management is with anticoagulation for thromboembolism, oxygen therapy, hydroxyurea and transfusion (6). We transfused our patient prior to starting her cesarean delivery and avoided using nitrous oxide.
- Acute fatty liver of pregnancy may cause coagulopathy, electrolyte abnormalities, multisystem organ dysfunction, hepatic failure, and perinatal mortality. Treatment is expedite delivery (7). We used GA with TIVA for the C/S after a brief amount of sevoflurane to minimize further liver damage associated with halogenated inhalational anesthetics and fatty liver of pregnancy.
- Our patient had a successful delivery with reasonably good post operative pain control because our multidisciplinary team were able to formulate and communicate different management plans based on the continually changing medical needs of our patient.



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