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Letter to Editor

# Perioperative Concerns in a Parturient with Hereditary Spherocytosis for Lower Segment Cesarean Section

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Hereditary spherocytosis (HS) is characterised by a spectrum of haemolytic conditions due to a red cell membrane defect. It has an autosomal dominant inheritance pattern, while variable expression of autosomal recessive pattern of inheritance may also be seen1. We hereby report the peri-operative management of a full- term obstetric patient with hereditary spherocytosis, presenting for elective lower segment cesarean section.

A 24-years -old female, known case of hereditary spherocytosis was posted for elective caesarean section. She gave history of yellowish discolouration of eyes since childhood. She was diagnosed with hereditary spherocytosis during her previous cesarean section two years back. There is past history of hospitalization at 17 years of age for blood transfusion due to severe anemia and abdominal pain. Examination during current hospitalization revealed icterus, with normal vital parameters. Systemic examination was unremarkable. Haemoglobin level of 10.4 was revealed on hemogram. The total leukocyte count and platelet counts were within normal range. Mean corpuscular volume (MCV) was found to be normal, but the reticulocyte count and osmotic fragility tests of red cells were elevated. Serum unconjugated bilirubin of 3.9 g/dl and conjugated of 1.3g/dl were noted. Renal function tests, serum electrolytes and other liver functions were within normal limits. Ultrasonography of the abdomen revealed hepatomegaly of 17cm and splenomegaly of 21cm (in addition to gravid uterus). Premedication in the form of intravenous ondansetron 4mg and intravenous pantoprazole 40mg were given. All standard ASA monitoring methods (electrocardiogram, pulse oximetry, non-invasive blood pressure cuff) were initiated. Under all aseptic precautions, subarachnoid block with patient in sitting position, at L3-4 interspace (Bupivacaine heavy 9 mg with 15 mcg fentanyl) was given and sensory block upto T6 level was achieved. A healthy 2.9 Kg baby with APGAR score of 10 was delivered. Intraoperative course was uneventful. Postoperatively, patient was hemodynamically stable and later shifted to a high-dependency unit(HDU) for observation and monitoring.

Hereditary spherocytosis is one of the familial haemolytic conditions associated with spherocytes in peripheral blood smear analysis. It is characterised by spherical red blood cells(RBCs) due to a defect in the membrane protein "spectrin", causing cytoskeletal instability2.These red cells with quantitative and qualitative abnormalities are more susceptible to lysis. Common complications include haemolytic crisis, aplastic crisis or megaloblastic crisis. Hemolytic crisis may be triggered by a viral infection, characterised by increase in jaundice, anemia, tenderness and increased enlargement of spleen. Diagnosis of HS may be made by classical haemolytic crisis or more often, gall stone disease. The diagnosis of HS in pregnancy is more complex as red cell fragility may be abnormal in pregnant women. Mild HS may be treated conservatively to keep haemoglobin above 9 g/dl, but recurrent haemolytic crisis may need splenectomy. Hemolytic crisis in pregnancy

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may be avoided by planned splenectomy in symptomatic HS patients before pregnancy. Splenectomy should be deferred until after delivery. More than half of these patients have pigmented gall stones3, increasing with age and severity of hemolysis. These patients may present to anaesthesiologists for various surgeries like cholecystectomy, splenectomy or any other surgery, including operative delivery. Perioperative management of HS predominantly depends on the degree of hemolysis and its consequences. For a major surgery, preoperative haemoglobin optimisation4 must be done along with arrangement of adequate blood and blood products. The perioperative insults which may contribute as triggers for crisis in HS should be avoided eg. hypoxia, hypercarbia, acidosis and hypothermia4. In our case, these issues were dealt meticulously in peri-operative period to achieve favourable outcome in the patient.

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