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EC ANAESTHESIA
Case Report

Anaesthetic Management of a Child with Hereditary Spherocytosis for Splenectomy

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Abstract

Hereditary spherocytosis (HS) is a familial heterogeneous disorder characterized by production of sphere shaped red blood cells (RBC) that are prone for chronic haemolysis resulting in haemolytic anaemia, splenomegaly, jaundice and cholelithiasis. The definitive diagnosis is made on history, clinical presentation, RBC Indices and genetic analysis. The anaesthetic management for splenectomy in children is challenging due to excessive RBC destruction related complications, liver dysfunction, acidosis and bleeding. There are certain perioperative concerns like anaemia, blood transfusion, hypoxemia and acidosis. Reporting anaesthetic management of nine years male child with hereditary spherocytosis with jaundice for splenectomy.

Keywords: *Hereditary Spherocytosis; Haemolysis; Splenomegaly; Splenectomy*

Abbreviations

HS: Hereditary Spherocytosis; RBC: Red Blood Cells; GA: General Anaesthesia; EA: Epidural Anaesthesia; ECG: Electrocardiogram; NIBP: Noninvasive Blood Pressure; SPO2: Oxygen Saturation; EtCO2: End Tidal Carbon Dioxide Concentration; Inj.: Injection; IV: Intravenous; PRBC: Packed Red Blood Cells; ETT: Endotracheal Tube

Introduction

The incidence of hereditary spherocytosis (HS) in Caucasian population is 1:2000 to 1:5000 which presents with mild to severe type of haemolysis [1]. In 80% of cases the inheritance of HS is autosomal dominant and in remaining as recessive genes or sporadic type. The normal concave shape of normal red blood cell (RBC) is replaced by spherical shape and is characterized by deficiency of ankyrin or spectrin which links the bilayer of red cells to the membrane skeleton resulting in increased susceptibility to lysis. An osmotic fragility test can help in the diagnosis of HS. Excessive haemolysis results in anaemia, jaundice, splenomegaly, cholelithiasis and alters the liver functions which affects metabolism of anaesthetic agents in these patients [2].

Case Report

Figure 1 a nine year old male child presented in outpatient department of paediatrics with fever, chills, vomiting and pain in abdomen associated with yellow discoloration of eyes and urine. Birth history of caesarean delivery for breech presentation weighing 2.8 kg, developed neonatal jaundice for which received phototherapy for 15 days. Milestones achieved, received vaccination up to date. H/O

hospitalization three months back under diagnosis of HS with hepatitis A, treated and discharged after 15 days. Child was referred for pre anaesthesia check-up for proposed splenectomy after five days. Family History- father also suffered from hereditary spherocytosis and underwent splenectomy at the age of 5 years.

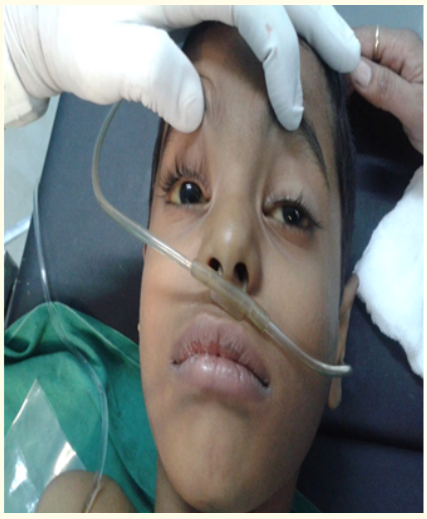

Figure 1: *Child with Icterus.*

On examination

Weight 20 kg (expected 30 kg), malnourished (grade II), active and afebrile. Pallor (+), Icterus (+) with no clubbing or oedema. Respiratory rate of 26/min, heart rate 92/min. On airway examination Malampatti class-I, all teeth present. On auscultation breath sounds and heart sounds were normal. On per abdominal examination tenderness in left hypochondriac region with palpable lump of size 6 x 8 cm on left side of upper abdomen.

Investigations

Hb: 13.3, TLC: 5300, Platelet count: 300,000, peripheral blood smear depicted spherocytosis and poikilocytosis. Incubated red blood cells showed increased osmotic fragility. Random blood sugar: 107, Urea: 16.8, Creatinine: 1.0 PT-INR: 15/14/1.0, Serum NH3: 120 (reference range: 30 - 80), Na+: 136, K+: 3.9, Cl-: 104.SGOT: 28.4, SGPT: 31.5, ALP: 250, Total Bilirubin: 27.6 mg/dL, direct bilirubin: 14.5 mg/dL, indirect bilirubin: 15mg/dL. Bleeding time: 2 minutes, clotting time: 5 minutes, HIV: nonreactive, HBs Ag: Negative. Ultrasonography of abdomen revealed splenomegaly. X-ray chest was normal.

Anaesthetic Management

After preoperative examination and optimization Informed risk consent was taken and general anaesthesia (GA) with epidural anaesthesia (EA) was planned. Two units of packed red blood cells and 1 unit of FFP was kept ready. The child was kept nil by mouth after 2 AM and taken for surgery at 9 AM. Preoperatively intravenous line (IV) was secured with 20 G IV cannula and ringer's lactate infusion started

5 ml/kg. Monitoring started for HR, NIBP, ECG, SPO_2, $EtCO_2$, temperature. IV premedication with Inj. emset 2 mg and Inj. glycopyrrolate 0.1 mg was given. The child was pre oxygenated with 100% FiO_2 with paediatric Bain's circuit and mask. Induced with Inj. Propofol- 20 mg IV. Intubation done with Portex 5.5 cuffed endotracheal tube (ETT) under the effect of Inj. choline 30 mg IV. Confirmed bilateral equal air entry and the cuff inflated, ETT fixed at 16 cm. Inj. vecuronium 3 mg given (loading dose). The child was given left lateral position and prepared for EA. Under aseptic precautions lumbar epidural space located with the help of paediatric 19G Tuohy's needle with loss of resistance technique at L_3 - L_4 spinal level. Epidural catheter of size 19G was inserted though the needle and fixed at 15 cm. Inj. bupivacaine 0.25% 10 ml given. Figure 2 child was positioned supine. GA maintained with 50% N_2O in O_2 + sevoflurane 1.5% + top-up doses of Inj. vecuronium 1 mg IV every 30 - 40 minutes (total 5mg). Inj. midazolam 1mg + Inj. fentanyl 25 mcg was supplemented IV intraoperatively. Epidural top up given with 5 ml 0.25% bupivacaine every 45 minutes. At the end of procedure all inhalational anaesthetic agents were stopped. After return of spontaneous ventilation and regaining of gag and cough reflexes, Inj. neostigmine 1 mg + Inj. glycopyrrolate 0.2 mg IV was given. The child was extubated after thorough oropharyngeal suctioning. Oxygenation given with face mask for next 15 min. Open splenectomy was performed by paediatric surgeon over two hours period. Figure 3 intraoperatively blood loss was approximately 150ml and urine output of 30 ml over 2 hours. PRBC 150 ml was transfused in the postoperative period. Intraoperative temperature was within range of 34.5^0C to 35.5^0C. The Intraoperative ABG as well as the hemodynamic were within normal limits throughout the surgery and recovery was uneventful. Figure 4 the child was shifted to recovery for further management with epidural catheter in situ to paediatric intensive care unit. Oxygen supplementation with 4 lit flow with nasal cannula was provided for 2 hours. For postoperative analgesia 5 ml of 0.2% ropivacaine was injected epidurally every 8 hours for 2 days and later epidural catheter was removed (Figure 5).

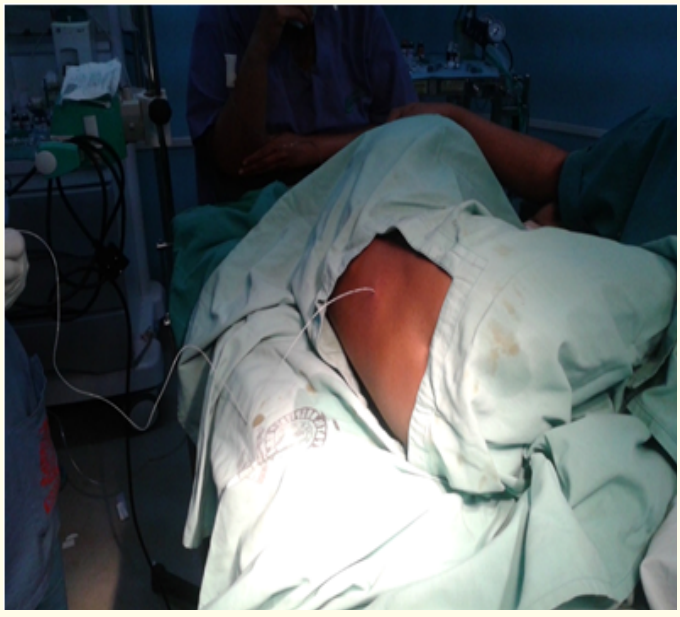

Figure 2: *Epidural Catheter in situ.*

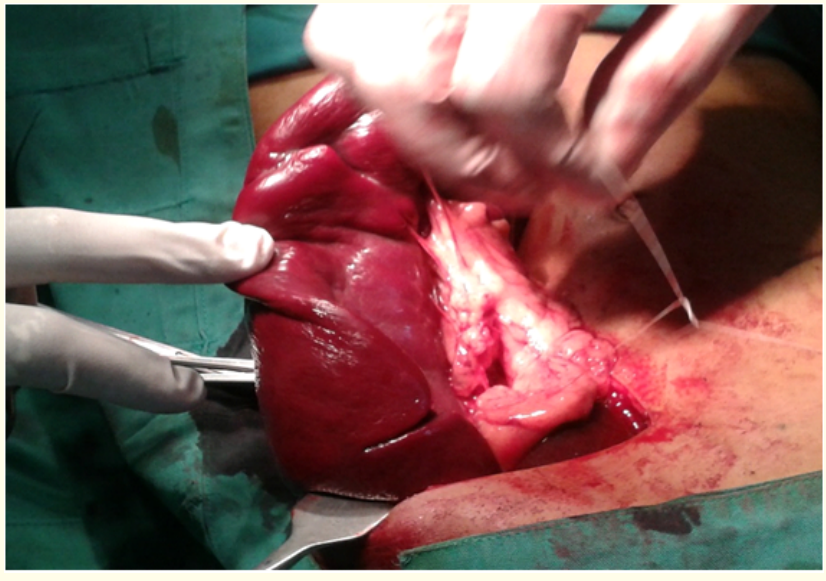

Figure 3: *Procedure of splenectomy.*

Figure 4: *Intraoperative hemodynamic parameters.*

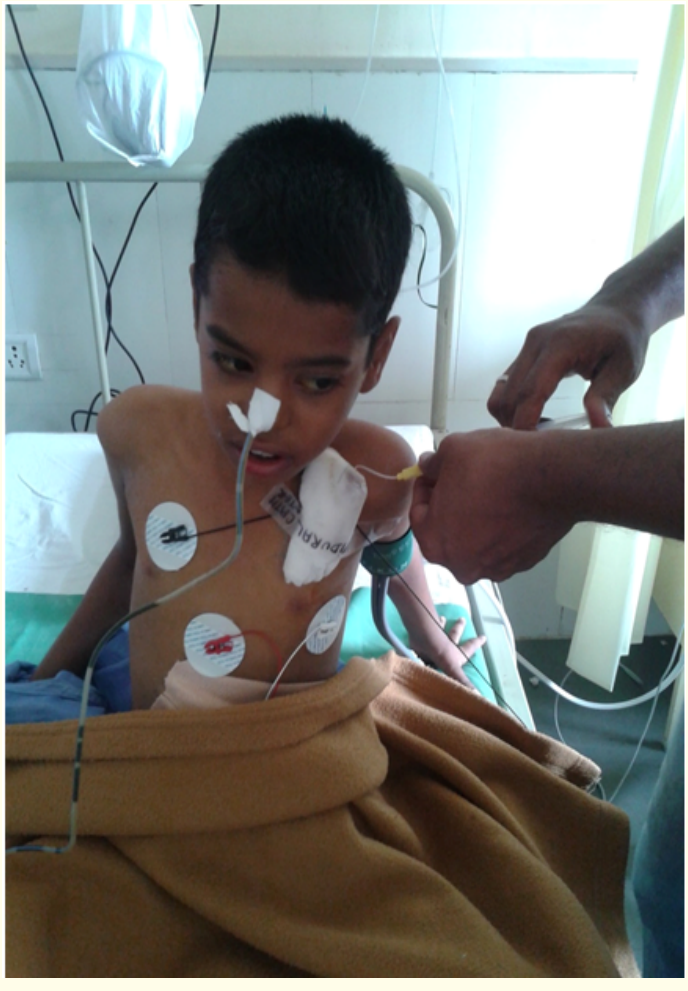

Figure 5: *Postoperative recovery.*

Discussion

Hereditary spherocytosis as autosomal dominant or recessive trait results due to genetic defects causing altered red blood cell membrane proteins that are responsible for normal biconcave shape of RBCs. The spherical shape loose flexibility and is liable for early destruction [1]. The spleen targets abnormal shaped RBCs but their passage into sinusoids is difficult due to spherical shape and gets phagocytosed resulting in extravascular haemolysis. In chronic cases infections or other illnesses cause frequent haemolytic crisis with anaemia, hypoxia, kernicterus and splenomegaly. Further indirect/direct bilirubin accumulates in gall bladder as gallstones, alteration in liver function and hepatomegaly develops [4]. Due to high RBC turnover with erythroid marrow activity children are vulnerable for the megaloblastic and aplastic crisis with parvovirus and other infections. Death can occur due to severe anaemia, heart failure and cardiovascular collapse [1].

Thus HS often presents in childhood with family history of the same. Splenectomy usually advised for moderate to severe condition in children with abdominal pain, anaemia that may be combined with cholecystectomy for gall stones. Splenectomy prevents further

haemolytic crisis, correction of jaundice and redevelopment of anaemia. The major pre-anaesthetic concerns are correction of anaemia, supplementation of folate to prevent megaloblastic crisis and immunization with *H. influenza* and pneumococcal vaccines. Intraoperatively adequate hydration to prevent stasis, avoidance of hepatotoxic drugs, prevention of hypoxia, acidosis assessed by ABG assessment, hypothermia prevention by administering warm fluids and proper room temperature and early replacement of blood loss with PRBCs-FFP transfusion. Postoperatively goals are oxygen supplementation, antibiotics, fluid and effective pain management.

We successfully managed this child with HS for splenectomy under combined GA with EA where we observed perioperative hemodynamic stability with smooth postoperative course as observed by Khatavkar., *et al.* [5] in an adult patient managed with GA+EA. GA with inhalational anaesthetic agents is known to reduce liver blood flow significantly but EA maintains the flow by decreasing stress response – release of catecholamines, pain and requirements of general anaesthetic agents.

Conclusion

Management of splenectomy for HS have many perioperative concerns in a child. But measures like correction of anaemia, immunization for infections, correction of blood loss and hypothermia. Vigilant intraoperative monitoring of the hemodynamic, temperature with avoidance/correction of acidosis, hypoxia and hypotension improves the surgical outcome and recovery.

Source of Support

Nil.

Conflict of Interest

None declared.

Acknowledgement

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