Anaesthetic management of beta thalassemia major in a child with hypersplenism posted for splenectomy

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Abstract
To study the anaesthetic management in a child with thalassemia major posted for splenectomy. The management of these children is challenging due to refractory anaemia, difficult airway, cardiomyopathy, pulmonary hypertension, intraoperative hypertension, restrictive lung diseases and postoperative infections.

Keywords- Beta thalassemia, splenectomy, anaesthesia management

INTRODUCTION

Thalassemia are hereditary disorders characterized by a reduction of synthesis of globin chain(alpha or beta)in haemoglobin molecule. Splenectomy is a common surgery performed in children of beta thalassemia major to reduce the frequency of transfusion and mechanical effect of enlarged spleen. Anaesthetic management of these children is challenging due to refractory anaemia, unanticipated difficult airway, perioperative high blood pressure, iron overload, endocrinological abnormalities, cardiomyopathies, restrictive lung disease , pulmonary hypoplasia and postoperative infections.

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Case report

A 7 year,8 months old child weighing 18kg born of nonconsanguineous marriage, diagnosed as beta thalassemia major at the age of 1 year was on multiple blood transfusion(every month) , was scheduled for splenectomy. child had received pneumococcal, meningococcal , hepatitis b ,influenza vaccine and last blood transfusion 2 weeks prior to surgery, on preoperative examination child had frontal bossing ,depressed nasal bridge, malar prominence and high arched palate.

Airway assessment was found normal Mallampati grade 3. He was pale ,icteric, not febrile with heart rate of 100 bpm. BP-110/7mmhg, Respiratory Rate of 24 with oxygen saturation of 98% on room air. No prominent neck veins, pedaledema. Systemic examination CVS- no murmurs, chest clear, hepatomegaly of 5 cm below right costal margin, Splenomegaly of 12 cm below left costal margin. hb-7g%,tc-9000/cm,pl-1.7lakhs.ferritin-1000 ng/ml (17-140 ng/ml)Lft-Tb-1.4,Db-0.6,IB-0.8SGOT-351 u/l,SGPT-352u/l. usg abdomen -gross splenomegaly

2 pints packed cells arranged for surgery

Anaesthetic management

Surgery was done under general anesthesia. iv line insitu, baseline parameters were normal.premedicated with midazolam ,fentanyl-50mcg,glycop. preoxygenated with 100%o2 for 3min,induced with propofol checked ventilation,paralysed with vecuroniumand intubated with 5.5mm endotracheal cuffed tube, fixed at 14cm,maintained with 02+n20+isoflurane/ boluses of vecuronium, meanwhile right internal jugular vein secured with 5.5F, 8 cm triple lumen cvc(8-10cmh2o) and right arterial line cannulated. Intraoperative vital signs stable,estimated blood loss was 400ml, replaced with 300 ml of packed rbc and ringer lactate, diclofenac suppository 25mg for postoperative analgesia, with return of adequate respiratory effort, neuromuscular blockade reversed with iv neostigmine and glycop, and extubated after thorough oral suctioning, and after the patient is awake, obeying commands

Discussion and conclusion:

Successful anaesthetic outcome of patients depends on properly planned airway and maintaining haemodynamic stability.

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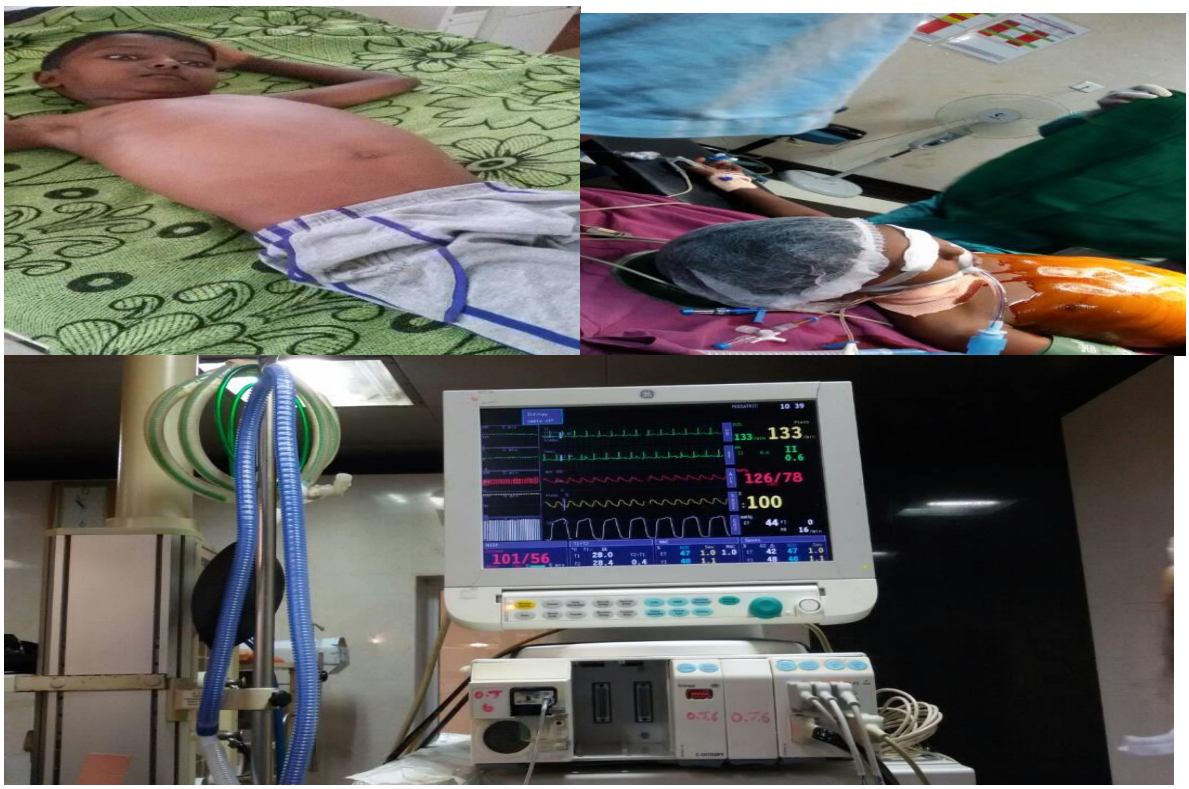

Intraoperative monitoring of heartrate, NIBP, IBP, CVP, SPO2