

# Anesthetic Management with Epidural Analgesia in a Case of Thalassemia Intermedia

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Thalassemia is an inherited defect in the production of globulin chains transmitted by autosomal recessive inheritance. Anaesthetic management of these patients is challenging, being associated with hypersplenism, unanticipated difficult airway and vertebral malformations, perioperative high blood pressure, systemic features of iron overload, cardiac and hepatic failure and anaemia. In this case, we highlight the anaesthetic concerns in a child of beta thalassemia intermedia with high levels of Hemoglobin F posted for splenectomy along with cholecystectomy. To avoid need of postoperative mechanical ventilation due to low oxygen carrying capacity of blood and anaerobic metabolism during the period of surgical stress, epidural catheter inserted despite possibility of epidural hematoma or bleed because of thrombocytopenia. Patient had an enhanced recovery in term of decrease oxygen demand, decrease postoperative nausea and vomiting, early ambulation and patient satisfaction. Intraoperative epidural also decreases need of neuromuscular blocking agent and intravenous analgesia. This lead to faster awakening from anaesthesia even in the presence of decrease oxygen carrying capacity due to anaemia and acidosis (anaerobic metabolism). Use of regional anesthesia should be considered in similar cases after assessing risk and benefits for individual cases.

**Keywords:** Thalassemia intermedia; Epidural anesthesia; Thrombocytopenia; Enhanced recovery

**T**halassemia is an inherited defect in the production of globulin chains transmitted by autosomal recessive inheritance. The final globin molecule is a tetramer of two alpha and two non- alpha chains. In beta thalassemia the clinical presentation typically manifests at four to six months of age, as during this time period haemoglobin F falls significantly, to be replaced by haemoglobin A [1-2]. They have symptoms attributable to anemia, hepatosplenomegaly, heart failure and skeletal changes secondary to bone marrow expansion. Anaesthetic management of these children is challenging being associated with unanticipated difficult airway, perioperative high blood pressure, iron overload, cardiac diseases and liver cirrhosis. Patients of thalassemia intermedia have chronic haemolytic anaemia but do not require transfusions except under periods of stress [3]. In this case, we highlight the anaesthetic concerns in a child of beta thalassemia intermedia with high levels of hemoglobin F posted for splenectomy along with cholecystectomy.

## Case Description

A 18-year-old male child weighing 28 kg known case of thalassemia intermedia was posted for elective open splenectomy and cholecystectomy. He was diagnosed with beta thalassemia when he was 6yrs old. In the past 1 year, he

received 17 transfusions until date of operation. Patient had complaints of abdominal distension and pain for the past 2 years. He was noted of growth diminution since 12 years of age with tanner stage 2 pubertal failure. On general examination, he had pallor, jaundice and tachycardia. Significant findings in cardiac assessment showed ejection systolic murmur over pericardium and abdominal examination showed marked hepato splenomegaly. Rest major systems were normal and no other skeletal abnormalities were evident on clinical examination. Patient was planned for splenectomy to reduce the need of blood transfusions along with cholecystectomy for gall bladder stones after getting him vaccinated.

Preoperatively airway was assessed and he had 3-finger mouth opening with grade 2 Mallampati grading and crowded teeth. Patient had a haemoglobin value of 6.9 gm/dl, haematocrit of 20.6%, platelet count of 90 000 per mm<sup>3</sup>, PT of 15.3 sec and INR of 1.35 with normal liver and renal function tests. Chest X-ray showed cardiomegaly with clear lung fields. ECG revealed left ventricular hypertrophy and sinus tachycardia. After application of the standard ASA monitors, a 19G epidural catheter was placed in T10-T11 space for perioperative analgesia in single attempt. Epidural infusion with 0.25% bupivacaine at 2ml/hour started after a bolus dose of 0.25% bupivacaine 10ml. Induction was achieved with propofol 50mg, fentanyl 50mcg and vecuronium 3mg. Patient was intubated successfully in single attempt (Cormack Lehane grade IIb) in spite of prominent malar eminence and protruding upper prominent incisors with 6.0 mm cuffed ETT. Patient was maintained on pressure-controlled ventilation with O<sub>2</sub>: N<sub>2</sub>O ratio of 50:50 along with sevoflurane. Invasive monitoring for goal directed fluid therapy was accomplished with an arterial line in left radial artery and right internal jugular vein central line

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(7FR B-BRAUN triple lumen catheter). Intraoperatively surgery proceeded for 3 hours with total blood loss around 800 ml and urine output 150 ml. Patient was transfused with four units of PRBC, 4 units of FFP and 4 units of platelet intraoperatively. After achieving hemodynamic stability and adequate reversal patient was extubated intraoperatively and was shifted to ICU for monitoring from where he was discharged to his ward uneventfully.

## Discussion

Anesthetic management in case of thalassemia depends on the severity of thalassemia and is a critical determinant of the postoperative morbidity. Marked medullary expansion, hepatosplenomegaly, growth retardation, intellectual impairment, facial anomalies, and hyperbilirubinemia may occur during the course of disease. Other symptoms like extra-medullary haematopoieses can occur in the vertebral canal, compressing the spinal cord causing neurologic symptoms as well as make performance of regional anesthesia difficult [4-5].

There are several anaesthetic implications of  $\beta$ -thalassaemia. In its milder form, only a chronic compensated anaemia is the major concern. With more severe forms, anaemia is severe and it is associated with feature of compensatory erythropoiesis. Frontal bossing, maxillary bone enlargement and crowning of teeth can make airway management difficult. If patients are on regular transfusions feature of iron overload (haemochromatosis) should be checked in organs like the heart (heart failure), endocrine system (endocrinopathy), and liver (cirrhosis) [6]. High incidence of blood transfusion related diseases like hepatitis can be present hence precautions must be taken to avoid exposure to blood and body fluids. Hypoxia, cardiovascular depression and hypovolemia should be avoided peri-operatively.

In one study of 100 thalassaemia children undergoing splenectomy, authors reported intraoperative and postoperative hypertension requiring aggressive treatment. The authors suggested manipulation of the large spleen during surgical removal resulting in auto transfusion as a possible cause of hypertension [7].

This case was representative of a more severe form of thalassemia intermedia with history of biweekly blood transfusions. He had decreased blood counts, features of heart failure, Cormack Lehane Grade IIb on intubation and his blood pressure also showed variable trends.

Upper abdominal surgery decreases functional residual capacity and splinting of diaphragm due to pain may compound problem of pulmonary atelectasis in postoperative period. To avoid need of postoperative mechanical ventilation we inserted epidural catheter despite being fully aware of possibility of epidural hematoma or bleed because of existing thrombocytopenia. Patient had an enhanced recovery in term of decrease oxygen demand,

decrease postoperative nausea and vomiting, early ambulation and patient satisfaction. Intraoperative epidural also decreases need of neuromuscular blocking agent and intravenous analgesia. This leads to faster awakening from anaesthesia even in the presence of decrease oxygen carrying capacity due to anaemia and acidosis (anaerobic metabolism). Use of regional anesthesia should be considered in such cases after assessing risk and benefits for individual cases. Studies reported very little chances of bleed when epidural anesthesia was given in patients with platelets count less than 100 000 mm<sup>-3</sup> [8]. Platelet transfusion can be done before performing regional anaesthesia, although there is no strong evidence to support it [9].

## Conclusion

In patients of Thalassemia, mortality and morbidity due to massive splenomegaly with hypersplenism, systemic organ failure and decrease oxygen capacity is significant. Benefits of a successfully placed epidural catheter include enhanced recovery after surgery. Anesthetic management require monitoring, early recognition and management of complications associated with thalassemia.

**Declaration of Patient Consent:** The patient has given his consent for his clinical information to be reported in the journal. The patient understands that his name and initials will not be published.

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