International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Anaesthetic Management for B Thalassemia Major Patient with Massive Splenomegaly for Splenectomy

Dr. Suhas H N[1], Deeksha P[2], Dr. Jyothsna[3]

[1, 2, 3]Department of Anaesthesiology and Critical Care, Ballari Medical Collage & Research Centre, Ballari

Abstract: ***A 14 yr old β Thalassemia Major patient with massive splenomegaly scheduled for elective splenectomy. Diagnosed at age 7, he exhibited easy fatigability, pallor, maxillary hyperplasia, and hepatomegaly. Initial hemoglobin was 7.9 g/dL increased to 8.7 g/dL post transfusion. Investigations showed elevated bilirubin and liver enzymes, with a peripheral smear indicating microcytic hypochromic anaemia. After obtaining informed consent, a 20G IV line was established, and monitoring commenced. General anaesthesia was induced with IV propofol and succinylcholine, followed by intubation with a 5.5 cuffed endotracheal tube. Anaesthesia was maintained with sevoflurane and titrated doses of atracurium and fentanyl. Intraoperative blood loss was 200 mL, with fluid replacement managed appropriately. Neuromuscular blockade was reversed, and the patient was extubated after suctioning. The postoperative period was uneventful, and discharge occurred on the 10th day. In conclusion, managing β Thalassemia Major requires careful preoperative assessment and readiness for complications.***

Keywords: β Thalassemia Major, splenomegaly, anaemia, blood transfusion, balanced anaesthesia

1. Introduction

β Thalassemia Major is an autosomal recessive genetic disorder predominantly affecting individuals of Mediterranean, Southeast Asian, Indian, and certain African descent, with an incidence estimated at approximately 1 in 100, 000 live births. This condition results from mutations in the HBB (haemoglobin subunit beta) gene, which encodes the beta - globin subunit of hemoglobin. The absence or reduction of beta - globin chains leads to an imbalance in globin production, resulting in ineffective erythropoiesis and a subsequent decrease in the synthesis of functional hemoglobin.

The hallmark clinical manifestations of β Thalassemia Major include severe microcytic hypochromic anaemia, characterized by reduced red blood cell (RBC) production and abnormal morphology. Patients often present with splenomegaly, as the spleen becomes engorged with the increased destruction of both maturing erythroblasts in the bone marrow and mature RBCs in the peripheral circulation. This hemolytic process contributes to the development of extramedullary hematopoiesis, which can further exacerbate splenomegaly.

Additionally, individuals with this disorder may experience significant bone deformities due to the expansion of the medullary cavity in response to ineffective erythropoiesis. Over time, this can lead to characteristic skeletal changes, such as maxillary hyperplasia and other skeletal abnormalities. The chronic nature of the anaemia also results in increased iron absorption from the gastrointestinal tract and frequent blood transfusions, leading to iron overload and its associated complications, including damage to vital organs such as the heart and liver.

Overall, the complexities of β Thalassemia Major necessitate a comprehensive management approach that includes regular blood transfusions, monitoring for iron overload, and potential surgical interventions such as splenectomy for patients with significant splenomegaly. Understanding the underlying pathophysiology is crucial for healthcare providers in delivering effective treatment and improving patient outcomes.

2. Case Report

History

14 year old male child of 25 kg weight, known case of HbE β Thalassemia Major diagnosed at age of 7yrs with massive splenomegaly scheduled for elective splenectomy. He was diagnosed at 7 years of age as HbE β Thalassemia Major & confirmed by HPLC with HbA - 54.1%, HbF - 16.30%, HbE - 29%

HbA	HbF	HbE
54.10%	16.30%	29%

HPLC- (old)

Osmotic fragility test - decreased suggestive of shift to left of RBC osmotic fragility. He had multiple blood transfusions & frequency of transfusion increased recently. Since one month his appetite had decreased & he complained of easy fatigability & reduced activity. Preoperatively he received one unit of PRBC transfusion, pneumococcal & meningococcal vaccination.

Clinical examination

On examination patient was conscious & oriented, had pallor with maxillary hyperplasia, pulse rate 108bpm, bp - 100/60mmhg, cvs –S1 S2 heard, no murmurs. His abdomen was distended and on palpation he had massive splenomegaly (extended upto right iliac fossa) & hepatomegaly**.**

Investigations

Upon investigating he had anaemia with haemoglobin 7.9 gm /dl initially which increased to 8.7 gm/dl after transfusing one

unit of PRBC and his coagulation profile was normal with prothrombin time 15 seconds, INR - 1.08. Liver function test were abnormal with elevated serum bilirubin & enzymes with normal renal function tests. Peripheral smear showed microcytic hypochromic anaemia & on ECG had sinus tachycardia and chest X - ray showed cardiomegaly. High risk consent was taken from parents & planned for general anaesthesia.

Anaesthetic management

In the operative room 20G intravenous line was secured, monitors were connected (includes pulse oximetry, NIBP, ECG, EtCO2, temperature) & baseline recordings noted and was premeditated with glycopyrolate 0.2 mg IV, midazolam 0.75 mg IV, fentanyl 50 mg IV & preoxygenated with 100 % oxygen for 3min and Induced with propofol 50 mg IV, suxamethonium 50mg IV, sevoflurane 2% oral intubation done with 5.5 cuffed endotracheal tube & bilateral air entry was confirmed and tube fixed at 16cm & connected to closed circuit. Anaesthesia was maintained with 02: AIR – 50%: 50% & 1% sevoflurane & titrated dose of atracurium & fentanyl.

Intraoperatively surgical blood loss was around 200 ml which was replaced with 150 ml PRBC & 500 ml of crystalloids, the rest of 200ml of PRBC was transfused postoperatively. Splenectomy was done & hemostasis achieved and Surgery lasted for 140 minutes. Intraoperatively hemodynamics was stable & throughout the surgery with HR - 80 - 120/min, systolic BP of around 90 - 110, diastolic BP of 60 - 70 mm hg, EtCO2 - 34 to 40mm of Hg, Spo2 - 99 to 100% & urine output was adequate. Residual neuromuscular blockade reversed with neostigmine 1.25 mg IV & glycopyrolate 0.2mg IV, once the child was awake & breathing regularly with adequate tidal volumes, thorough oral suction done & extubated then child shifted to post anaesthetic care unit for observation & postoperative period were also uneventful & child was discharged on 10th postoperative day.

3. Discussion

Thalassemia Major is a severe hemoglobinopathy caused by absent or decreased synthesis of one of the globin chains, primarily affecting the production of beta - globin. This condition results in a significant reduction in the concentration of hemoglobin A (HbA), accompanied by an increase in the levels of hemoglobin A2 (HbA2) and fetal hemoglobin (HbF). The imbalance in globin chain production leads to microcytic hypochromic anemia, characterized by small and pale red blood cells, as well as a relative excess of unpaired globin chains. These excess chains precipitate as insoluble inclusions within erythrocytes, which ultimately contributes to their premature destruction.

In β Thalassemia Major, the ineffective erythropoiesis occurs primarily due to the inability of the bone marrow to produce functional red blood cells in adequate numbers. This results in the destruction of maturing erythroblasts within the marrow, leading to ineffective erythropoiesis, while mature red blood cells are lysed in the spleen, contributing to hemolysis. The resulting anaemia triggers an increase in erythropoietin production as the body attempts to stimulate red blood cell production. However, due to the underlying pathophysiology of the disease, these compensatory mechanisms are largely ineffective.

The clinical presentation of Thalassemia Major is severe and often includes pallor, icterus (jaundice), and splenomegaly. In the case of the patient discussed, he presented with severe microcytic hypochromic anaemia and massive splenomegaly, along with abnormal liver function tests indicating potential hepatic involvement due to hemolysis and iron overload.

A peripheral blood smear in patients with Thalassemia Major typically reveals notable features, including hypochromic microcytic red blood cells, anisocytosis (variation in red blood cell size), poikilocytosis (abnormal red blood cell shapes), target cells, and basophilic stippling. Fragmented red blood cells are also common, reflecting the hemolytic process. The reticulocyte count may be elevated, indicating a compensatory response to anemia.

Chronic hemolytic anaemia leads to secondary complications such as hepatosplenomegaly, leg ulcers, gallstones, and high - output cardiac failure due to increased workload on the heart. Children with untreated Thalassemia Major often experience profound growth retardation, increased susceptibility to infections, and endocrine dysfunction, significantly impacting their quality of life and leading to early mortality.

Regular blood transfusions are a critical component of management for these patients, improving anaemia and suppressing the secondary erythropoietic features. However, the necessity of repeated transfusions introduces the risk of iron overload, potentially resulting in secondary hemochromatosis. This iron overload can lead to severe complications, including damage to vital organs such as the heart, liver, and endocrine glands.

In this specific case, the child experienced failure to thrive, easy fatigability and reduced activity levels, with a diagnosis of HbE β Thalassemia Major established at the age of 7. He had undergone repeated blood transfusions for the past six years to manage his anaemia. This long history of transfusion dependency underscores the importance of careful monitoring for iron overload and related complications, alongside addressing the underlying disease through comprehensive management strategies.

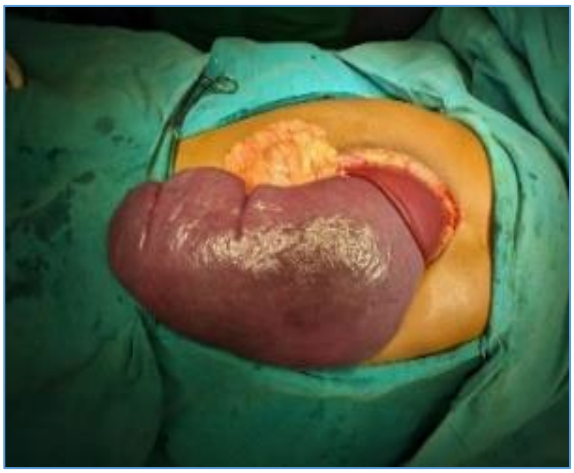
Overall, the complexities of Thalassemia Major require an integrated approach, combining transfusion therapy, iron chelation, and supportive care, to optimize patient outcomes and enhance quality of life. Awareness of the clinical manifestations and potential complications is crucial for healthcare providers to deliver effective care and interventions.

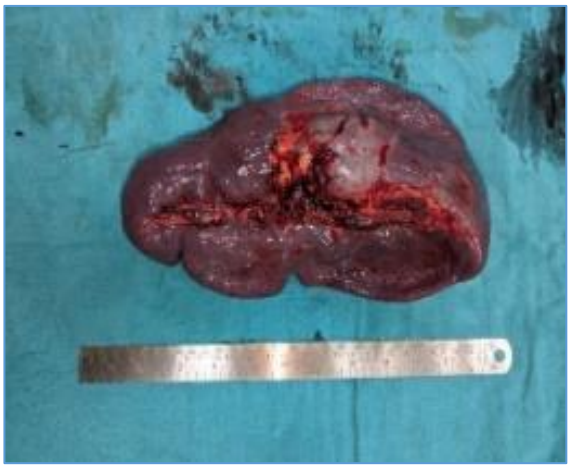
Concerns:

Anaemia, Altered LFTs, intraoperative bleeding, difficult airway due to bone marrow expansion & medullary erythropoiesis leading to skull & facial deformities (thalassemia facies - chipmunk facies), hepatosplenomegaly, leg ulcer, gallstone, high output cardiac failure, growth retardation susceptible to infection, endocrine dysfunction & die at early age are the major anaesthetic concerns.

Patients with β Thalassemia Major often present with anaemia and may have varying degrees of fluid overload or dehydration. Preoperative assessment of volume status is essential, as it guides fluid resuscitation strategies. During splenectomy, significant intraoperative fluid shifts can occur due to factors such as blood loss, manipulation of the spleen, and changes in vascular resistance. Rapid shifts from intravascular to extravascular compartments may lead to hypotension and decreased organ perfusion. Therefore, Continuous monitoring of hemodynamic parameters, urine output, and fluid balance is crucial throughout the surgical procedure. Invasive monitoring (such as arterial lines) may be warranted to provide real - time data on blood pressure and cardiac function, allowing for timely adjustments in fluid therapy. Avoiding intraoperative hypoxia, hypercarbia, and hypothermia in patients with β Thalassemia Major undergoing splenectomy is crucial for ensuring patient safety and optimal outcomes.

Repeated blood transfusions required to improve anaemia & suppress secondary features related to erythropoiesis results in iron overload & secondary hemochromatosis. Adequate preparation, balanced anaesthesia, intraoperative blood transfusion with intense monitoring helped in successful outcome in current case.





4. Conclusion

β Thalassemia Major includes various anaesthetic problems intraoperatively like difficult intubation, bleeding complication, hypothermia, hypoxia, hypercarbia, which were managed successfully. Hence adequate knowledge on pathophysiology, clinical features & complications of β Thalassemia & appropriate preparedness are important in successful management of these patients undergoing consequent surgeries including splenectomy

References

[1] Jyothi B, Sushma KS, Syeda S, Raza SO. Anaesthetic management of beta thalassemia major with hypersplenism for splenectomy in pediatric age group: Report of four cases. Anesth Essays Res 2015; 9: 266 - 9. Source
[2] T Kallenbach (2015) Anaesthesia for a patient with beta thalassaemia major, Southern African Journal of Anaesthesia and Analgesia, 21: 5, 140 - 143, DOI: 10.1080/22201181.2015.1076108
[3] Firth PG. Anesthesia and hemoglobinopathies. Anesthesiol Clin.2009 Jun; 27 (2): 321 - 36. doi: 10.1016/j. anclin.2009.05.001. PMID: 19703679.
[4] Gupta N, Kaur S, Goila A, Pawar M. Anaesthetic management of a patient with Eisenmenger syndrome and β - thalassemia major for splenectomy. Indian J Anaesth.2011 Mar; 55 (2): 187 - 9. doi: 10.4103/0019 - 5049.79892. PMID: 21712881; PMCID: PMC3106397