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Anesthetic Management for Splenectomy in Pediatric Patient with Beta Thalassemia Major: A Case Report

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Abstract

Thalassemia is a genetic disorder of the hemoglobin protein in red blood cells.
Thalassemia major is the homozygous type with severe degree of symptoms; thalassemia minor is the heterozygous type with decrease severity of symptoms [1]
Patients with this disorder may require multiple transfusions therefore splenectomy is a common surgery performed in this group of patients to decrease the frequency of transfusion and mechanical effect of the enlarged spleen. Anesthetic management in these patients are potentially challenging because of the unanticipated difficult airway, associated cardiac diseases, restrictive respiratory pattern, increased risk of perioperative hypertension, underlying endocrinological abnormalities, iron overload state and greater risk of pulmonary hypertension.[2]
This is case report describes the anesthetic management in a 17 year old female with Beta thalassemia major undergoing elective splenectomy

Keywords: Beta Thalassemia, genetic disorder, hemoglobin, multiple blood transfusion, splenectomy

Introduction

Thalassemia is a genetic disorder of the hemoglobin protein in red blood cells. It has been classified into thalassemia minor, intermedia and major, depending on the genetic defect and severity of the disease. [3]The clinical presentation of β-thalassemia varies widely from a mild asymptomatic form in thalassemia minor, to a severe disease in thalassemia major where individuals are dependent on life-long blood transfusions.[4] The hallmark of thalassemia syndromes is the production of defective red blood cells that are removed by the spleen resulting in an enlarged hyperfunctioning spleen (splenomegaly).[5] Annual transfusion volume exceeding 225 to 250 mL/kg per year with packed red blood cells (hematocrit 75 percent) may indicate the presence of hypersplenism.[6] Splenectomy is indicated in the transfusion-dependent patient with hypersplenism. It will prolong the red blood cell survival by reducing the amount of red blood cells removed from circulation and may ultimately result in the reduced need for blood transfusions.[7]

This case report describes the anesthetic management in a patient with massive splenomegaly suffering from Transfusion dependent Beta thalassemia major undergoing therapeutic splenectomy.

Case Report:

A 17-Year-old female, weighing 30 kgs, suffering from thalassemia major diagnosed at the age of 6 months, presented with chief complaints of fever for 2 days, bilateral pedal edema for 1 day and mass per abdomen which was gradually progressive in nature for two years. Her history revealed that she was on multiple blood transfusions once a month depending upon the hemoglobin requirement. Patient was

prophylactically on iron chelator T. Deferasirox 250 mg once a day.

On examination, she had frontal bossing, malar prominence and high arched palate. Airway assessment was found normal and Mallampati grade 1 with bucking of teeth. She was pale, afebrile with a heart rate (HR) of 112 bpm, Blood pressure (BP) of 116/58 mmHg, respiratory rate of 34 cycles/min, Lung fields clear with air entry bilaterally equal, S1, S2 heard with raised JVP and associated gallop present. normal oxygen saturation of (SpO2) 98% on room air. Bilateral Pedal edema was present until the knee.

Per abdomen examination revealed prominent splenomegaly 10 cm below left costal margin. Ultrasonography showed a spleen of span 15.1cm with normal echotexture.

On admission her hemoglobin (Hb) was 3gm/dl without any overt signs and symptoms of bleeding. Patient was admitted in the PICU and was transfused packed cell volume according to 20cc/kg over 10 days. Patient was given Inj Lasix at 0.5ml twice a day for 1 day in view of heart failure signs. Patient was kept on oxygen support through a non-rebreather mask.

After optimum stabilization and management. The pre operative hematological investigations revealed Hb of 9.7 gm/dl and platelet count of 91,000/cu.mm after six units of packed cell transfusion. A preoperative echocardiography was performed which relieved an LVEF of 60% with trivial MR, Trivial PR, Trivial TR along with grade IV diastolic dysfunction. Pulmonary arterial pressure was 26mmHg with pulmonary artery diastolic pressure of 13 mmHg. Signs of left ventricular volume overload seen on echocardiography.

Rest all hematological parameters were within normal limits. She had received pneumococcal, haemophilus influenza and meningococcal vaccinations prior to surgery. After assessment an elective open Splenectomy was planned by the surgeon in view of massive splenomegaly and frequent blood transfusion. Surgery was performed under general anesthesia; she was kept nil per mouth eight hours for solids and two hours for clear liquids prior to surgery. Premedicated the night prior with tablet Pantoprazole 20 mg and tablet alprazolam 0.25 mg.

On the day of the surgery after the preoperative assessment a Peripheral venous access was secured with a 22-gauze cannula.

Upon arrival into the operating room, patient was laid supine, monitors were attached and vitals like non-invasive blood pressure (NIBP), electrocardiogram (ECG) and oxygen saturation (SPO2) was recorded.

The Baseline HR was 130 bpm, NIBP 112/76 mmHg, SPO 2 98% on room air and respiratory rate (RR) 18 cycles/min. Preoxygenation was done with 100% oxygen. Premedicated with Injection midazolam 0.05 mg/kg and fentanyl 2 µg/kg.

Induction was done with Injection propofol 2 mg/kg, after ensuring appropriate ventilation. Injection vecuronium 0.1 mg/kg was given, the patient was ventilated and the airway was secured with a cuffed endotracheal tube (ETT) of size 6.5 mm.

Bilateral air entry checked it was found to be equal and adequate, and thus the tube was fixed at 20 cm depth. Considering major surgical procedure would be performed additional analgesia was planned but due to low platelet count epidural analgesia could not be given. Thus, the patient was given USG guided Transversus abdominis plane block on the right side with 15 ml of 0.2% Ropivacaine to combat the intraoperative and postoperative pain management.

Under anesthesia invasive arterial access secured with 4 FR radial arterial line on right side and Intra Arterial blood pressure was monitored along with other vital monitoring.

Volume 7, Issue 3; May-June 2024; Page No 539-544

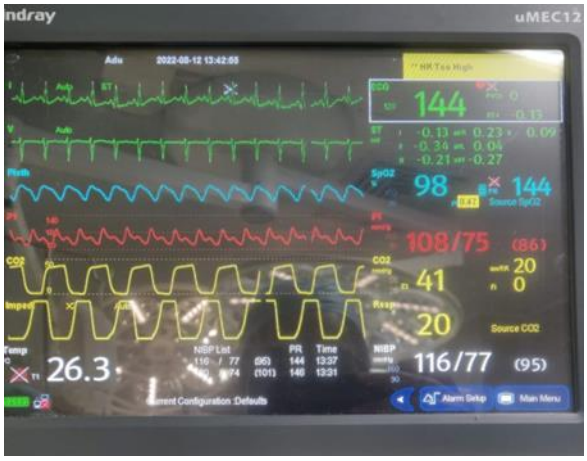


Fig. No. 1 : Intraoperative vitals

Fig. No. 2: Intraoperative Intubated image

On the request of surgeon nasogastric tube was inserted without any difficulty and secured for decompression of stomach. (Fig. No. 2).

Surgery started with a left subcostal incision. Intraoperatively during the procedure, the patient's saturation showed fluctuation. During evaluation coarse crepitations were heard on auscultation in bilateral upper lobes of the lung and was suspected to have pulmonary edema. Patient was given 2 puffs of Salbutamol each containing 100mcg directly into the endotracheal tube along with adequate suctioning. This was followed by Injection Dexamethasone 6 mg along with Injection Hydrocortisone 60 mg.

Patient additionally received a bolus of 60 mg Inj Furosemide in divided dose to combat the signs of pulmonary edema. Despite this extensive management, the patient still had audible coarse crepitations thus the patient was started on an infusion of Injection Furosemide 200 mg in 50 ml normal saline at the rate of 6 ml/hr to deal with going pulmonary edema.

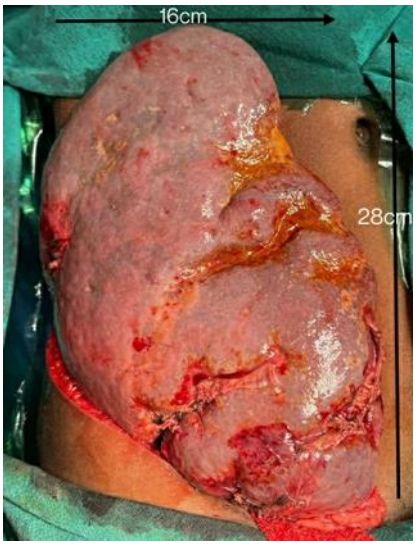
Following this the patient started developing hypotension and thus Injection Ketamine 20 mg was given since it's a potent bronchodilator and will increase blood pressure without having any effect on pulmonary blood pressure.

Volume 7, Issue 3; May-June 2024; Page No 539-544

Rest of the intraoperative period was uneventful with stable hemodynamic parameters and no adventitious sounds.

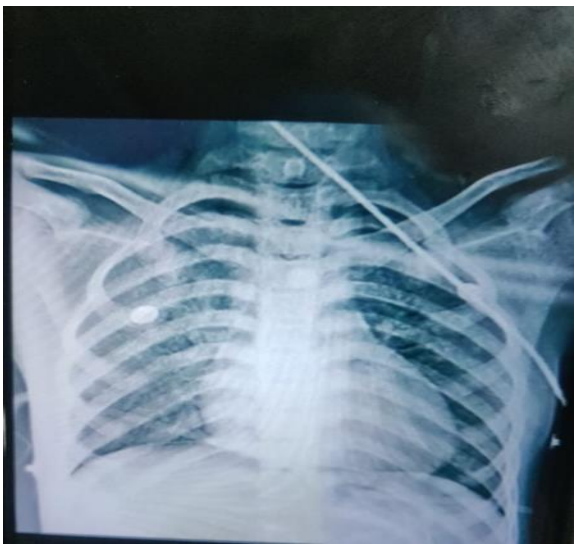
The patient was given 100 ml ringer lactate solution intraoperatively with urine output of 1050 ml and blood loss of 250 ml. The Surgery lasted for two and half hours.

Fig. No. 3 Spleen specimen



Prior to extubation an ABG was performed which was within normal limits and chest auscultation showed no audible crepitations. After meeting up the extubation criteria, when the patient's spontaneous respiratory efforts were equal and continuous, a trial of extubation was given and neuromuscular blockade was reversed with Injection Neostigmine 0.05 mg/iv and Injection Glycopyrrolate 0.008 mg/iv. Patient was successfully extubated. ETT was removed and the patient was awake and obeying commands. 450 mg Injection paracetamol was given for postoperative analgesia. Patient was shifted to PICU for observation and monitoring with a continuous infusion of Inj Furosemide 200mg in 50ml normal saline at 6ml/hr. Postoperative period was uneventful and infusion was stopped 2 hours after the surgery. Patient was maintained on room air in the ICU with stable hemodynamic parameters.

Fig. No. 4 : Post operative Xray



Volume 7, Issue 3; May-June 2024; Page No 539-544

Patient was kept in the ICU for 4 days and during the stay in the ICU patient was transfused PRBC @10ml/kg on post operative day 3 in view of low hemoglobin (Hb- 7.6g/dl) . Patient was shifted to wards on the 5th post operative day and then discharged 12 days later from the surgery. The hematological parameter on discharge was Hb 10.4 g/dl and Platelet 2.35/cumm on discharge.

Discussion

Thalassemia affects millions of individuals across the globe, not many people are aware about the condition thus World thalassemia day is celebrated on 8th May in order to create awareness regarding the disorder.[8]

The β thalassemias pose a significant health burden in India. The average prevalence of β thalassemia carriers is 3–4% which translates to 35 to 45 million carriers in our multi-ethnic and culturally and linguistically diverse population of 1.21 billion people which also includes around 8% of tribal groups according to the Census of India 2011.[9] Several ethnic groups have a much higher prevalence (4–17%). Estimates indicate that there would be around 100,000 patients with a β thalassemia syndrome in this vast country. 4 The prevalence of pathological hemoglobinopathies in India is 1.2 per 1000 live births. It has been suggested that there would be 32,400 babies with a serious haemoglobin disorder born each year based on 27 million births per year in India. 5 Of the 10,000 to 12,000 thalassemic children born annually in India, very few are optimally managed mainly in urban regions.[10]

Thalassemia is an autosomal recessive disorder. It is characterized by impaired production of normal globin chains (alpha and beta) resulting in excess of one type of chain. Beta thalassemia is characterized by impaired synthesis of the beta chain, whereas alpha thalassemia results from impaired synthesis of the alpha chain. [11] This impaired globin chain synthesis results in intravascular hemolysis, extramedullary hematopoiesis, erythroid hyperplasia, profound anaemia, severe bone deformities, hepatospleno-megaly, growth retardation and death by second or third decade.[12] Pulmonary hypertension and progressive vascular damage occur because of chronic hemolysis and disturbed nitric oxide physiology. Maxillary bone enlargement due to extramedullary hematopoiesis results in airway difficulty. Preoperative evaluation include examination of organs like heart, liver, spleen and endocrine system (pancreas and pituitary) affected by hemochromatosis following multiple transfusions.[13] Management includes supportive treatment like multiple blood transfusions. Splenectomy is indicated only when there is splenomegaly or treatment of transfusion related iron overload. Allogeneic bone marrow (stem cell) transplantation can cure severely affected patients.[14] Prevention of Thalassemia is a public health concern and cost effective when compared to treatment.This can be done by preimplantation genetic diagnosis and noninvasive prenatal diagnosis centres. Aggressive screening of a pregnant woman can bring down the burden of this disease. [15 ,16]

Various obstacles observed include management of difficulty in securing a patent airway because of extramedullary hematopoiesis. Anemia which persists in spite of transfusion which can result in reduced oxygen carrying capacity of the blood causing deranged intraoperative hemodynamic parameters. Pulmonary hypertension and intra operative systemic hypertension can result in severe V/Q mismatch due to restrictive lung disease which is caused by hypersplenism and decreased oxygen carrying capacity. There is a high incidence of blood transfusion related diseases like hepatitis hence precaution must be taken to avoid exposure to blood and body fluids.[17] In our patient, presence of thalassemic facies made the airway management typically challenging. Our patient also received iron chelation therapy, which is actually beneficial in preventing cardiac dysfunction and reducing liver iron concentrations. The FiO 2 was kept 70% or above throughout the surgery, to avoid any V/Q mismatch and pulmonary complications. Immunization with pneumococcal,haemophilus influenza and meningococcal vaccinations prior to surgery were given to prevent post operative infections. Postoperative follow-up of patients revealed there was improvement in Hb% and decreased requirements of blood transfusions.

Conclusion

Thalassemia is a common form of haemoglobinopathy in developing countries. It is a challenge for surgeon as well as anesthetist because of difficult airway owning to the presence of thalassemia facies and deranged hemodynamic parameters and increased risk of intraoperative and perioperative cardiac and

Volume 7, Issue 3; May-June 2024; Page No 539-544

respiratory complications. However, all these perioperative risk were managed effectively with preoperative planning and intraoperative vigilance.

References

1. Uranus S, Sill H. Splenectomy for hematological disorders. In:Holzheimer RGMannick JA, editors. Surgical Treatment: Evidence based and Problem-oriented. Munich: Zuckschwerdt; 2001
2. Firth PG. Anesthesia and hemoglobinopathies. Anesthesiol Clin. 2009;27:321–36.
3. Cohen AR, Galanello R, Pennell DJ, Cunningham MJ, Vichinsky E. Thalassemia. ASH Education Program Book. 2004 Jan 1;2004(1):14-34.
4. Cao A, Galanello R. Beta-thalassemia. Genetics in medicine. 2010 Feb 1;12(2):61-76.
5. Galanello R, Origa R. Beta-thalassemia. Orphanet journal of rare diseases. 2010 Dec;5:1-5.
6. Lal A, Wong TE, Andrews J, Balasa VV, Chung JH, Forester CM, Ikeda AK, Keel SB, Pagano MB, Puthenveetil G, Shah SJ. Transfusion practices and complications in thalassemia. Transfusion. 2018 Dec;58(12):2826-35.
7. Smith CH, Erlandson ME, Stern G, Schulman I. The role of splenectomy in the management of thalassemia. Blood. 1960 Feb 1;15(2):197-211.
8. Aydinok Y. Thalassemia. Hematology. 2012 Apr 1;17(sup1):s28-31.
9. Yadav SS, Panchal P, Menon KC. Prevalence and management of β-thalassemia in India. Hemoglobin. 2022 Jan 2;46(1):27-32.
10. Colah RB, Gorakshakar A. Control of thalassemia in India. Thalassemia Reports. 2014 Sep 29;4(2):1955.
11. Muncie Jr HL, Campbell JS. Alpha and beta thalassemia. American family physician. 2009 Aug 15;80(4):339-44.
12. Origa R. β-Thalassemia. Genetics in Medicine. 2017 Jun;19(6):609-19.
13. Javad G, Saeid A, Mohammadmehdi N. Thalassemia and immune system dysfunction-review article. Int J Curr Res. 2011 Sep 26;3:105-8.
14. Borgna-Pignatti C, Gamberini MR. Complications of thalassemia major and their treatment. Expert review of hematology. 2011 Jun 1;4(3):353-66.
15. Fucharoen S, Winichagoon P. Prevention and control of thalassemia in Asia.
16. Mendiratta SL, Mittal M, Naaz F, Singh S, Anand S. Role of thalassemia screening in prevention and control of thalassemia--a 5 year experience. International Journal of Reproduction, Contraception, Obstetrics and Gynecology. 2016 Sep 1;5(9):3107-12.
17. Bharati S, Das S, Majee P, Mandal S. Anesthetic management of a patient with sickle β+ thalassemia. Saudi Journal of Anaesthesia. 2011 Jan 1;5(1):98-100.
18. Cappellini MD, Cohen A, Eleftheriou A, Piga A, Porter J, Taher A. Blood transfusion therapy in β-thalassaemia major. InGuidelines for the Clinical Management of Thalassaemia [Internet] 2nd Revised edition 2008. Thalassaemia International Federation.
19. N. Madan, S. Sharma, S.K. Sood, R. Colah, H.M. Bhatia Frequency of thalassemia trait and other hemoglobinopathies in northern and western India Indian J Hum Genet, 16 (2010), pp. 16-25
20. I.C. Verma, R. Saxena, S. Kohli Past, present and future scenario of thalassemic care and control in India Indian J Med Res, 134 (2011), pp. 507-521.
21. K. Grow, M. Vashist, P. Abrol, S. Sharma, R. Yadav β thalassemia in India: current status and challenges ahead.
22. Schrier SL, Angelucci E. New strategies in the treatment of the thalassemias. Annu Rev Med. 2005;56:157–71.
23. Roshan Colah, Khushnooma Italia, Ajit Gorakshakar, Burden of thalassemia in India: The road map for control, Pediatric Hematology Oncology Journal, Volume 2, Issue 4 ,2017.

Volume 7, Issue 3; May-June 2024; Page No 539-544