

Case Report

Anaesthetic management of two patients with β -thalassaemia intermedia

Shahnawaz Ali, Fauzia Anis Khan

Department of Anaesthesia, Aga Khan University, Stadium Road, Karachi, Pakistan.

Abstract

There is paucity of literature regarding the anaesthetic management of patients with thalassaemia intermedia. In this case study, the anaesthetic management and concerns in two children with thalassaemia intermedia aged eleven and nine years undergoing herniotomy and splenectomy respectively is reported. Both children had unanticipated difficulty in airway management and high intraoperative blood pressure trends. These cases are representative of the range of problems seen in children with thalassaemia intermedia.

Introduction

Thalassaemias are hereditary disorders characterized by reduction in the synthesis of globin chains (α or β) in the haemoglobin (Hb) molecules. This reduction in synthesis of haemoglobin produces hypochromic microcytic anaemia. Normal adult haemoglobin is primarily haemoglobin A (HbA). In β -thalassaemia, reduced β -globin chain synthesis results in a relative increase in the percentages of Hb A₂ and F compared to Hb A. Patients homozygous for a milder form of β -thalassaemia have thalassaemia intermedia. Genotypically they have Hb A value of 0-30%, Hb A₂ 0-10% and Hb F 6-100%. These patients have chronic haemolytic anaemia but do not require transfusions except under periods of stress. If transfusion requirement is high they may also develop iron overload. They survive into adult life but with hepatosplenomegaly and bony deformities.¹

Literature on anaesthetic management of thalassaemia exists but we were unable to locate any specific case reports of anaesthetic management of children with thalassaemia intermedia. We report the management of two cases with β -thalassaemia intermedia and review of literature on the anaesthetic concerns associated with this disease.

Case Reports

An eleven years old female child presented for bilateral inguinal hernia repair. On preoperative examination the patient had frontal bossing and tonsillar enlargement. Her airway was assessed and found normal. Her haemoglobin and haematocrit were 9.1 gm.dl⁻¹ and 28.1% respectively. Hb A₂ was 2.4% and Hb F was 97.6%. She had never been transfused.

Midazolam 5mg was used as premedication. Monitoring included pulse oximeter, non-invasive blood

pressure (NIBP), ECG and capnography (EtCO₂). Patient was induced with sevoflurane and oxygen. Pethidine 0.8 mg.kg⁻¹ was used as an analgesic. An attempt was made to insert size 2 laryngeal mask airway (LMA) but difficulty in placement was encountered due to a high arched palate. Patient's trachea was then intubated with a size 5.5 mm endotracheal tube after administration of atracurium. Intubation grade was Modified Cormack and Lehane IIb. Anaesthesia was maintained with O₂:N₂O 1:1, isoflurane at 1 MAC (Minimum Alveolar Concentration) and bolus doses of atracurium. EtCO₂ was within normal limits. Baseline heart rate was 120 beats.minute⁻¹ and it was maintained between 120-130 beats.minute⁻¹, blood pressures showed a trend towards hypertension intra-operatively with systolic blood pressure around 130 mmHg (baseline 90 mmHg) and diastolic blood pressure 80-90 (baseline 40) mmHg. Anaesthesia and surgery lasted for 90 minutes. Two hundred milliliters of crystalloid solution was administered. Recovery and postoperative periods were uneventful.

Second patient was a nine years old male child, a known case of thalassaemia intermedia who presented for elective splenectomy. Splenectomy was scheduled to reduce blood transfusion requirements as patient was being transfused with one unit of packed cell every month for last four years. Preoperatively airway was assessed as normal. Patient had a haemoglobin value of 8.7 gm.dl⁻¹, haematocrit of 25.1%, with HbA₂ 1.5% and HbF 98.5%. ECG revealed a heart rate of 91 beats.minute⁻¹, and left ventricular hypertrophy with T wave inversion in V1-V3.

Child was premedicated with midazolam. Monitors used were pulse oximeter, noninvasive blood pressure (NIBP), ECG and capnograph. Induction was achieved with thiopentone 5mg.kg⁻¹, fentanyl 2ug.kg⁻¹ and atracurium 0.5mg.kg⁻¹. On direct laryngoscopy, Cormack Lehane grade was found to be IIb. Intubation was done with the help of a stylet. Patient was then ventilated with O₂:N₂O ratio of 1:1. EtCO₂ was maintained within normal limits. Baseline heart rate was 100 beats.minute⁻¹ and was maintained between 100-115 beats.minute⁻¹. Blood pressures in the beginning were 90/40 mm Hg and remained in the range of 100-110 mm Hg systolic blood pressure and 60-70 mm Hg diastolic blood pressure in spite of analgesia. Blood loss was minimal. Surgery lasted for 150 minutes. Three hundred and fifty milliliters of crystalloid solution was administered.

Extubation and postoperative periods were uneventful.

Discussion

β -thalassaemia primarily affect persons from Mediterranean origin and to a lesser extent Chinese, other Asians, and blacks.¹ In Pakistan, thalassaemia is the most prevalent genetically transmitted blood disorder with a carrier rate of 5-8%; around 5000 children are diagnosed each year in the country.²

Kitoh et al reported the management of an adult patient with thalassaemia intermedia and haemolytic crisis that underwent splenectomy.³ Patient had hyperkinetic left ventricular motion and increased cardiac index. This patient was successfully managed using isoflurane at low concentration and fentanyl. In another case report successful management of an adult patient with thalassaemia intermedia and severe haemolytic anaemia undergoing emergency splenectomy was discussed.⁴

Patients with β -thalassaemia intermedia generally do not require blood transfusions. But if the presentation of the disease is severe and these patients are not adequately transfused; marked medullary expansion, hepatosplenomegaly, growth retardation, facial anomalies, and hyperbilirubinaemia may occur.⁵ Extra-medullary haematopoieses can occur in the vertebral canal, compressing the spinal cord and causing neurologic symptoms. These patients may present with nutritional haemosideroses (if transfused) and other complications of thalassaemia.⁶

There are several anaesthetic implications of β -thalassaemia.⁷ Patients with β -thalassaemia intermedia can also present with these difficulties if the disease is severe and/or the treatment is delayed. Extra medullary haematopoieses resulting in maxillary bone enlargement can make airway management difficult. If patients are on regular transfusions, preoperative evaluation should focus on those organs most affected by haemochromatosis: the heart, endocrine system, and liver. There is also a high incidence of blood transfusion related diseases like hepatitis; hence precautions must be taken to avoid exposure to blood and body fluids. The incidence of RBC alloimmunization in children with thalassaemia is approximately 5%, making provision of compatible blood a potential problem. Preoperative transfusion to adequate levels should be performed. Hypoxia, cardiovascular depression and haemoglobinuria should be avoided peri-operatively.

In our cases, the first child was not transfusion dependent and had no specific features of blood transfusions and thalassaemic children except for anaemia and frontal bossing. The problems encountered were difficulty in LMA placement and a slight difficulty in tracheal tube insertion along with trend towards intra-operative hypertension. The

second case was representative of a more severe form of thalassaemia intermedia had a history of monthly blood transfusions for last four years along with frontal and maxillary bossing. He did have abnormal liver function tests and signs of myocardial strain. He also had Cormack and Lehane IIb intubation and his blood pressure also showed a trend towards higher side. Variations in facial bone anatomy in thalassaemic children is thought to be the cause of difficult airway in these children. These two cases elicit the variation in symptoms and signs observed in this disease of which anaesthetists need to be aware about.

One retrospective study of 100 thalassaemic children undergoing splenectomy reported intraoperative and postoperative hypertension requiring aggressive treatment. The authors suggested manipulation of the large spleen during surgical removal resulting in autotransfusion as a possible cause of hypertension.⁸ In another study of thalassaemic children undergoing splenectomy, furosemide decreased the incidence of postoperative convulsions but had no significant effect on the incidence of postoperative hypertension.⁹

Slight difficulty in managing the airway can be explained by facial anomalies that develop in this group of patients. Repetitive blood transfusions throughout the life and autotransfusion during splenectomy are considered as possible causes of increased blood pressure. We were unable to find a satisfactory explanation for isolated intraoperative hypertension in both cases despite analgesia and adequate depth of anaesthesia. There is scarcity of information about this group of thalassaemic patients undergoing anaesthesia and surgery, and more published literature is needed to make further guidelines.

Acknowledgement

We would like to thank Dr. Salman Naseem Adil, Associate Professor and Head of Haematology and Oncology Section, Aga Khan University, Karachi, for his suggestions in improving this case report.

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