



Anaesthetic Management of a Child with Factor IX Deficiency Planned for Adenotonsillectomy

Deepali Gupta^{1*}, Dhruv Gupta², Ashish Aneja³, Radha Panchapakesan⁴, Arulmurugan Balasubramanian⁵, Taha M Abdelaal⁶, Mohamed Othman⁷, Samia Al Saddik⁸, Steffi Paulson John⁸ and Youmna Ayman Faheem⁸

¹Anesthesiologist in Nmc royal hospital, Khalifa city, UAE

²Head Dermatologist and HOD in Ahalia Hospital, Abudhabi, UAE

³Physician in yas clinic Abudhabi, UAE

⁴Anesthesiologist in Burjeel hospital, Abudhabi, UAE

⁵HOD Anesthesia Phoenix Hospital, Abudhabi, UAE

⁶ENT Consultant NMC Royal hospital, Abudhabi, UAE

⁷Anesthesia consultant in nmc royal, UAE

⁸Intern at Nmc Royal Hospital, Abudhabi, UAE

*Corresponding author: Deepali Gupta, Anaesthesiologist in Nmc royal hospital, Khalifa city, UAE; E-mail: drdeepali30@hotmail.com

Abstract

Kids with bleeding disorders going for adenotonsillectomy needs meticulous advance planning to avoid any complications. The main complication from adenotonsillectomy for children with bleeding disorders is bleeding. Extra precautions to manage bleeding must be taken before, during and after the surgery.

Keywords: Hemophilia; Adenotonsillectomy; Haemorrhage

Introduction

The risk of haemorrhage post adenotonsillectomy surgery is 0.2 to 2.2 for a primary haemorrhage, and 0.1 to 4.8 for a secondary haemorrhage. These surgeries are done frequently in the paediatric population and bleeding is one of the most common postoperative complications a patient can encounter. This paper describes a case of a five year old male patient with a history of Factor IX deficiency who presented to the hospital for his adenotonsillectomy surgery as planned. In addition, in his preoperative tests it was found that his labs were suggestive of an underlying coagulation disorder. The tests ordered were his prothrombin time (PT) and activated partial thromboplastin time (aPTT) activities in the plasma. However, in this case; it was a good decision to perform these tests as the patient would have suffered from severe haemorrhage during and after the surgery. Preoperative testing should be done in order to ensure that each patient is safe before, during and after the surgery. Depending on the severity of the disease, factor IX concentrate may be given

prior to dental extractions and surgery to prevent bleeding. The prognosis is usually good with treatment.

Background

The main feature of haemophilia B, also known as Christmas disease. Haemophilia is a hereditary disorder which results from deficiency of one of the blood's clotting factors. The X chromosome contains both the factor VIII and factor IX genes. Haemophilia B results from factor IX deficiency; it is inherited through an X-linked recessive pattern, predominantly affecting males, but in rare cases could also affect females. Haemophilia B affects 1 in 40,000 live males, or about 15% of haemophilia cases, it is less common than haemophilia A, which affects 1 in 5,000 live males, or about 85% of haemophilia cases, and haemophilia C affects 1 in 100,000 live births, or about 1% of haemophilia cases. All races and ethnic groups are equally affected. Consanguinity is a major factor in increasing frequencies in certain communities, such as Egyptians and Ashkenazi Jews. Individuals with severe haemophilia B are usually diagnosed shortly after birth or within the first 1-2 years of life; those with

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moderate haemophilia B, are diagnosed at five to six years of age; and those with mild haemophilia B, may find out in adulthood. Some people have mild disease, some moderate, and some more severe. Patients with more serious cases of haemophilia often get regular shots of the factor that they're missing — known as clotting factor replacement therapy — to prevent bleeding episodes. The clotting factors are transfused through an intravenous (IV) line, and can be given in the hospital, at the doctor's office, or at home.

Case Presentation

Five year old male patient presented to the ENT clinic with a history of sore throat, breathing difficulties and recurrent tonsil infections, and was planned for an intracapsular tonsillectomy and adenoidectomy done under general anaesthesia. However, prior to the surgery he was diagnosed with moderate haemophilia B as per his preoperative lab assessment and past history of prolonged bleeding after a circumcision surgery at five months of age. The patient is currently not on any chronic medications, no history of any medical illnesses or a similar complaint in the past. In addition, there is no significant family history with regards to any bleeding disorder. Prior to his surgery, coagulation tests were done as per routine assessment, and it was found that his bleeding time was prolonged—also known as partial thromboplastin time (PTT). Therefore, this warranted further evaluation which showed that the patient had some deficiency in one or more of his blood tests. These deficiencies included a low Factor IX activity, a low Factor VIII activity, and a low Von Willbrand activity as well. Therefore, he was diagnosed with a mild haemophilia B defect. The patient was then treated with tranexamic acid 500 mg tablets for two weeks, and was instructed to take his first dose one day prior to the surgery in order to avoid any complications before, during, or after the procedure. Moreover, 25 mg/kg PO, tranexamic acid is indicated in patients with haemophilia for short-term use (i.e, 2-8 days) to reduce/prevent haemorrhage and reduce the need for replacement therapy. In addition, since this is a moderate case of haemophilia B, the patient was not transfused with Factor IX before surgery. The child was planned for general anaesthesia (GA) and had minimal side effects. Prior to the surgery, the child was pre-assessed by an anaesthesiologist for past medical history, any drug history and allergies. Moreover, the child's airway, heart, and lungs were also examined by the anaesthesiologist to confirm the absence of any airway problem such as wheezing and coughing. After the assessment was completed, the patient was posted for the procedure under GA. In addition, some blood work up was done, the patient was diagnosed with a Factor 8 deficiency and preparations were done accordingly— such as the availability of Factor 8 in the operation theatre (OT) in case of haemorrhage. Anaesthesia for adenotonsillectomy is a skill test for the anaesthesiologist. Our

Aim is to provide safe anaesthesia by avoiding any complications and to bring a successful outcome. Premedication was given which included oral midazolam 6 mg twenty minutes before going for surgery in order to decrease the patient's anxiety. This helps the child to separate from the parents smoothly and not remember the experience. General anaesthesia for tonsillectomy and adenoidectomy usually begins by having your child breathe aesthetic gas through a mask. Sevoflurane 6% was used for induction, and the child was pre-oxygenated for three minutes with 100% oxygen. Once the child was asleep, an intravenous (IV) line was secured with a 22 G cannula. The following medications were given for GA Intravenously; Propofol 30 mg IV Injection, Fentanyl 20 mg IV injection, Rocuronium 10 mg IV injection. Moreover, oral intubation was done with a number five endotracheal tube (ETT) as the child weighed 20 kgs, and was fixed at 14.5 cm. Finally a ventilator was connected. The child was antagonized with Neostigmine 50 micrograms per kg and Atropine 20 micrograms per kg. Throughout the surgery, the patient was maintained on oxygen nitrous 50.50, and Sevoflurane 2%. As a result, the surgery went smoothly and there were no intraoperative complications like bleeding or fluctuating vitals. The patient was monitored closely, keeping an eye out on the ECG, pulse, temperature, Oxygen and CO2 levels and neuromuscular monitoring. Intra-operatively, paracetamol 15 mg/kg was given, along with Ringer's Lactate ½ DNS 150 mL IV. In addition, dexamethasone 2 mg IV was also given with Ondansetron 1.5 mg IV. Tranexamic acid 100 mg was also infused via IV for management of postoperative haemorrhage and other complications. There were no complications like bleeding and hypotension during or after the surgery. The endotracheal tube was removed and the child was extubated in the head low and left lateral position (also known as the post tonsillectomy position). The throat was freed of any secretions and suctioned under vision. The patient was then shifted to the recovery room in the lateral position and was closely monitored for bleeding or any deterioration in consciousness. The recovery process was good and the child's temperature, ECG, pulse, and SpO2 were monitored. Postoperatively, the following analgesia was advised; Pethidine 10 mg IV for effective pain control, Ondansetron 0.1 mg/kg IV for nausea control, and dexamethasone 0.15 mg/kg IV in case the child complains of pain and nausea in the postoperative period. The child was then shifted to the ward after monitoring for 6 hours in the post anaesthesia care unit. After which he stayed in the ward for 24 hours for observation of any postoperative bleeding, fever or related complications because the patient has a history of a bleeding disorder. His parents were advised to continue following up with the paediatric haematologist and ENT doctors [1-13].

Follow-Up

Postoperatively, the patient was taken to the anaesthesia care unit and was sought after for any bleeding, or complications. Patient followed up in the clinic two weeks later with the ENT doctor,

and then with the paediatric haematologist as well. Finally, there were no signs of postoperative bleeding, or infection (Tables 1,2).

Table 1: Investigations.

LAB TEST	DATE	RESULT	RANGE
FACTOR VIII	7 DECEMBER 2022	52 % (LOW)	57-163
FACTOR IX	7 DECEMBER 2022	15 % (LOW)	52-111
FACTOR X	7 DECEMBER 2022	113 % (NORMAL)	76-183
FACTOR XI	7 DECEMBER 2022	74 % (NORMAL)	60-150
FACTOR XII	7 DECEMBER 2022	102 % (NORMAL)	50-150
aPTT	7 DECEMBER 2022	34.9 seconds (borderline)	26.0-35.0
Lupus Anticoagulant	7 DECEMBER 2022	40.5 seconds	0.0-44.0
vWF Activity	7 DECEMBER 2022	41 % (LOW)	50-200
vWF Antigen	7 DECEMBER 2022	61 % (NORMAL)	50-200

Table 2: Treatment.

Medicine Name	Strengt h	Dosage	Frequen cy	Duration	Start Date	End Date	Quantity	Instructions
TRANEXA MIC ACID 500 MG TABLETS	750 mg	750	TID	1 week	26/12/2022	1-1-2023	21	Start one day before operation

Discussion

The presence of coagulation disorder in children who are operated for adenotonsillectomy is a challenge for the Surgeon and Anaesthetist. Adenotonsillectomy can be done safely in patients with haemophilia B, but the increased risk of postoperative haemorrhage cannot be ruled out. Haemophilia B is the most common hereditary coagulopathy and is due to a quantitative and/or qualitative deficiency of Factor IX. Proper preoperative history and physical examination with emphasis on past bleeding and family history can decrease the complications. Children undergo laboratory screening and Hematologic evaluation to determine the presence of a hereditary or acquired coagulopathy. This patient was diagnosed with Factor IX deficiency. Patients are diagnosed to have haemophilia B if they had one or more abnormal values of factor IX, vWF antigen, or aPTT, along with a family or patient bleeding history or two or more abnormal laboratory values in the absence of a family or bleeding history. Our child had low factor VIII, IX, low vWF activity with prolonged aPTT. Positive history of bleeding was there during his circumcision surgery. The management plan included proper screening, tranexamic acid and extra doses of factor IX surrounding the surgery.

Conclusion

We conclude that patients with bleeding disorders undergoing adenotonsillectomy are at a higher risk of bleeding and require

close monitoring with haemostatic support for a prolonged period of time in post-operative period.

Learning Points

Haemophilia B is a bleeding disorder due to Factor IX deficiency, and most commonly affects male patients—rarely females. Bleeding is the main complication from adenotonsillectomy for haemophiliac children. Therefore, the paediatric haematologist and otolaryngologist (ear, nose and throat, or ENT specialist) should coordinate a treatment plan in order to prevent any adverse events in future surgeries.

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