

ANAESTHESIA FOR INCIDENTAL SURGERY IN A PATIENT WITH EBSTEIN'S ANOMALY

Dr. Rathna¹ Dr. Tejesh C. A.² Dr. Manjunath A. C.³ Dr. Mathews K. Thomas⁴

SUMMARY

Ebstein's anomaly accounts for <1% of congenital heart disease and is associated with varied clinical manifestation and unique haemodynamic derangements. We present a case of Ebstein's anomaly who underwent a successful open appendectomy under general anaesthesia.

Keywords : *Ebstein's anomaly, Anaesthetic management, General anaesthesia.*

Introduction

Ebstein's anomaly is a rare congenital cardiac abnormality, wherein, the septal and posterior cusps of the tricuspid valve are displaced downwards and are elongated, such that varying amount of right ventricle effectively forms part of right atrium.¹ Most patients also have an interatrial communication (ASD, patent foramen ovale) through which there may be right-to-left shunting of blood.² They carry risk of tachyarrhythmia and profound hypoxemia during anaesthesia.³

Case report

A 45yr old male with recurrent appendicitis and incidentally detected to have Ebstein's anomaly was referred to our hospital for open appendectomy. Patient presented with recurrent episodes of right lower abdominal pain for the past 6 months. He had no history of dyspnoea on exertion and his effort tolerance was good to moderate. There was no history of chest pain, palpitations, syncope, or swelling of the lower limbs. On examination, pulse rate was 76/min, regular, good volume; BP was 126/84 mmHg in right upper limb, no cyanosis, no pedal edema and JVP was not raised. Cardiovascular examination revealed a grade II pansystolic murmur in tricuspid area. Patient's Hb% and PCV was 14.9 mg% and 45% respectively. ECG showed first degree heart block and RBBB. Echocardiographic examination showed Ebstein's anomaly with dilated right atrium, grade II tricuspid regurgitation and ejection fraction of 50%.

Patient was premedicated with alprazolam 0.25 mg the night before surgery, pantoprazole 40 mg and ondansetron 4 mg on the night before surgery and in early morning. Infective endocarditis prophylaxis was given. In the OR after securing intravenous access with an 18 G cannula, midazolam 1.5 mg and fentanyl 100 µg was given intravenously. NIBP, ECG and pulse oximetry monitors were instituted. CVP was not instituted as it carries risk of arrhythmias in these patients.⁴

Anaesthesia was induced with propofol 2 mgkg⁻¹ I.V. given titrated to loss of eye lash reflex, as these patients have prolonged induction times.⁵ After induction, relaxation was achieved with vecuronium 0.1 mgkg⁻¹ I.V. and patient was intubated with No.8.5 oral endotracheal tube. Anaesthesia was maintained with oxygen, nitrous oxide and isoflurane 0.6-0.8% on circle system with controlled ventilation. Patient was haemodynamically stable throughout the intraoperative period with MAP between 84-100 mmHg and heart rate between 60-80/min. Intraoperatively ETCO₂ was ranging from 28-32 mmHg and SpO₂ 98-99%. Surgery lasted for duration of 40 minutes. At the end of surgery, when the patient had respiratory attempts, neuromuscular blockade was reversed with neostigmine 2.5 mg and glycopyrrolate 0.5 mg. Recovery and extubation was smooth and uneventful.

During the first 24 hrs postoperatively patient was monitored in ICU. Postoperative period was uneventful and the patient was discharged home on 7th postoperative day.

Discussion

Ebstein's anomaly is a rare congenital cardiac abnormality accounting for <1% of congenital heart disease.⁶ The septal and posterior cusps of the tricuspid valve are displaced downwards. Hence, varying amount of right ventricle effectively forms part of the atrium (atrialization of ventricle). The remaining functional part of the right ventricle is therefore small.¹ The tricuspid valve is usually

1. M.D., Prof. & Head
Dept. of Anaesthesiology & Critical Care,
MS Ramaiah Medical Teaching Hospital, Bangalore-54.
 2. M.D., Lecturer
 3. M.D., Lecturer
 4. M.D., Asst. Prof.
- Correspond to :**
Dr. Tejesh C. A.
E-mail : drtejeshca@yahoo.com

regurgitant, but may also be stenotic. Most patients have an interatrial communication through which there may be right-to-left shunting of blood.²

The degree of abnormality of right ventricular function and size of the ASD are the main determinants of the severity of the condition, which varies considerably. The natural history of the disease is very variable. 50% of case present in neonates and infants with cyanosis and congestive heart failure. In those who survive to adulthood, symptoms may be precipitated by the onset of arrhythmias, or by pregnancy. A few patients remain asymptomatic even in adulthood, in whom the anomaly is discovered incidentally.⁷

Preoperative abnormalities¹

- There may be a right-to-left shunt, with dyspnoea and cyanosis at rest, or on moderate exertion. Alternatively, the patient may be asymptomatic.
- Episodes of tachyarrhythmias occur in 25% of patients. Some provoke syncopal attacks.
- ECG may show, large peaked P waves, prolonged PR interval, WPW syndrome and RBBB. Paroxysmal supraventricular tachycardia occurs because of presence of WPW syndrome.
- Chest X ray may show cardiomegaly, with a prominent right heart border and poorly perfused lung fields.
- Paradoxical embolization, bacterial endocarditis, brain abscess and congestive heart failure may occur.
- A number of other lesions of the tricuspid valve or right ventricle may mimic Ebstein's anomaly, therefore the discriminating clinical and echocardiographic features for correct diagnosis have been enumerated.⁸

Anaesthetic problems¹

- Induction times are prolonged because of pooling of drugs in the large atrial chamber.⁴
- Intracardiac catheter insertion may be hazardous because it can provoke serious arrhythmias.^{1,5}
- Air entering peripheral venous lines or any open veins at subatmospheric pressure may cause paradoxical air emboli.
- Tachycardia is poorly tolerated because of impaired filling of the functionally small right ventricle.
- Hypotension may increase the right-to-left shunt.
- Hypoxia causes pulmonary vasoconstriction, which also increases right-to-left shunt.

- There is risk of bacterial endocarditis, especially if a CVP line is in place.
- Deterioration may occur with onset of arrhythmias and in pregnancy.

Management

- Severity of lesion must be assessed clinically and echocardiographically.
- Heart failure and arrhythmias require treatment.
- Antibiotic prophylaxis against bacterial endocarditis.
- If CVP is used for monitoring, its tip should be kept within the superior venacava.
- Although CVP monitoring or insertion of a pulmonary artery catheter may be used to evaluate cardiac preload, these measures are technically difficult in Ebstein's anomaly and lead to complications such as tachyarrhythmias or paradoxical emboli.
- TEE is useful to monitor cardiac output.
- Techniques should aim to minimize tachycardia and hypotension.
- Aim for cardiovascular stability – etomidate, propofol, fentanyl or alfentanil and vecuronium are suitable.

General anaesthesia has several advantages over regional technique in patients with Ebstein's anomaly. Hypotension is usually avoided and although this complication can be treated with fluids and vasopressors, both these may result in further instability in these patients. Excessive administration of fluid may lead to congestive heart failure or may increase right-to-left shunt and hypoxemia. Ephedrine is a potent beta agonist which may precipitate supraventricular tachycardia in these patients. General anaesthesia seldom causes hypotension and endotracheal intubation enables control of oxygen delivery.

Takafumi Horishita et al³ reported a case of Ebstein's anomaly wherein, a 58 yr old patient underwent left ankle fracture under epidural anaesthesia. The authors used echocardiography for intraoperative evaluation of cardiac preload and cardiac function. They avoided CVP monitoring or pulmonary artery catheter insertion as it may lead to complications such as tachyarrhythmias and paradoxical emboli. Linter SP and Clarke⁹ successfully employed epidural anaesthesia for elective cesarean delivery. Epidural anaesthesia may be appropriate in non-severe patients, but excessive administration of fluid should be avoided because it may increase right atrial pressure sufficiently to cause an increased right-to-left shunt and hypoxemia. Grover VK

et al⁴ accomplished an abdominal operation under general anaesthesia. They used a peripherally inserted catheter through the axillary vein to monitor central venous pressure. The patient developed bacterial endocarditis postoperatively despite antibiotic prophylaxis.

A search in the past journals reveals a paucity of literature about the anaesthetic management of patients with this rare congenital cardiac abnormality coming for incidental surgeries.

In conclusion, meticulous planning, understanding of haemodynamic derangements, individualisation and titration of drugs to the point of requirement and diligent care even for the minor deviations are an essential requisite for successful outcome of such rare congenital cardiac abnormalities.

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