

Case report

Anesthetic Management of a Patient with Ebstein's Anomaly for Obstructed Inguinal Hernia

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Abstract

Ebstein's anomaly is a rare congenital heart disease constituting less than 1% of patients with congenital heart diseases. It has diverse presentations, severity, and consequently, the management too. These patients are at increased risk of developing ventricular and supraventricular tachycardia, congestive heart failure, and death. Here, we report a case of a 32-year older man with Ebstein's anomaly, who had successfully undergone emergency open inguinal hernioplasty under nerve blocks.

Keywords: Ebstein's anomaly, Non-cardiac surgery, Nerve blocks, Obstructed inguinal hernia

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Introduction

Ebstein's anomaly (EA) is a congenital cardiac defect accounting for <1% of congenital cardiac anomalies (1). It is not only a rare but also a complex and fascinating con with various anatomical and clinical presentations. It was named after Wilhelm Ebstein, who first described the heart of a 19-year-old patient named Joseph Prescher. EA has the following features: 1. Apical displacement of the septal and posterior tricuspid valve (TV) leaflets. 2. Atrialization of the right ventricle (RV). The severity of the condition depends on the extent of valvular pathology, intracardiac shunting with patent foramen ovale, pulmonary hypertension, and associated tachyarrhythmia or Wolf-Parkinson-White (WPW) syndrome (2). Perioperative management of the patient with this cardiac anomaly for a non-cardiac surgery

poses significant challenges to the anesthesiologists. A few case reports are available in the literature regarding the anesthetic management of patients with EA for non-cardiac surgeries (2-6). However, to the best of our knowledge, the administration of a case under nerve blocks for inguinal hernioplasty has been reported. Here, we report a case of a 32-year-old man, a known case of EA, who had successfully undergone emergency open left inguinal hernioplasty under nerve blocks. We have obtained consent from the patient for publishing the case details in any journal for academic purposes.

Case Report

A thirty-five years old man, with a diagnosis of obstructed left inguinal hernia, was posted for

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emergency left inguinal hernioplasty. He was a known case of Ebstein's anomaly for 10 years and was on digoxin and beta-blockers for 2 years. He had exertional dyspnea of New York Heart Association Class 2 and palpitations. He had no history of recurrent chest infections and chest pain and no history suggestive of cyanosis. General examination revealed grade 2 clubbing, pedal edema, and raised jugular venous pulsation. His blood pressure was 120/80; heart rate 86/min, SpO₂ 100% on room air, and no respiratory system abnormalities clinically. Cardiovascular examination revealed parasternal heave, raised JVP, and on auscultation, S₁-widely split, S₂ was standard, and a holosystolic murmur over the tricuspid area was heard. The patient had no other co-morbidities and no other congenital heart diseases. Blood investigations were regular. ECG showed right bundle branch block (RBBB), steady rhythm, and right ventricular hypertrophy with right axis deviation. Chest X-ray revealed cardiomegaly. Echocardiography showed - Ebstein's anomaly of TV, grossly dilated right atrium, arterialization of RV, septal tricuspid leaflet shifted towards RV, tricuspid regurgitation, no evidence of atrial septal defect, normal LV systolic function. Ultrasound of the abdomen reported dilatation of inferior vena cava and hepatic vein, minimal right-sided pleural effusion, and mild ascites.

Infective endocarditis prophylaxis was given as per the cardiologist's opinion. Proper precautions were taken to avoid air bubbles in the peripheral venous lines. We opted for ilioinguinal and iliohypogastric nerve blocks if the patient obtained informed consent explaining the merits and demerits of all anesthetic techniques.

In the operating room, monitors including an electrocardiogram, pulse oximeter, and non-invasive blood pressure were attached. The patient was sedated with midazolam 2mg and fentanyl 100mcg intravenously. Under aseptic precautions, ilioinguinal and iliohypogastric nerves were identified between the internal oblique and transverse abdominal muscle with ultrasound guidance (Sonoscape, Linear probe 5-10 MHz) using an out-of-plane approach and a total of 15 ml of 0.5% bupivacaine was injected until its spread was confirmed. After ensuring the success of the blocks by the pin-prick method, surgery was

commenced. The surgeon was requested to infiltrate the sac with the local anesthetic for complete surgical anesthesia. Hemodynamics were stable during the procedure, which lasted for 70 minutes. The post-operative period was uneventful, and the patient was discharged home on the 4th post-operative day.

Discussion

EA is a rare congenital cardiac lesion, with less than 1% incidence among congenital heart diseases, and was first described by Wilhelm Ebstein in 1866 (1). It is characterized by dysplastic abnormalities of both basal and free attachments of the tricuspid valve leaflets, with downward displacement and elongation of the septal and anterior cusp, resulting in tricuspid regurgitation and rarely stenosis. Therefore, varying amounts of the right ventricle from the part of the atrium (arterialization of the ventricle) result in a small functional right ventricle. In addition, inter-atrial communication through a patent foramen ovale might also be associated with this condition resulting in the right to left shunting (1). The anticipated complications are cyanosis, pulmonary hypertension, cardiac arrhythmias, pulmonary and systemic emboli, congestive cardiac failure, and sudden death (6).

A few case reports are available in the literature for anesthetic management of patients with EA for non-cardiac surgeries under different techniques (2-6). Continuous epidural, single-dose caudal epidural, GA was successfully administered in pregnant patients with EA (2, 5, 6), while GA was safely administered for laparoscopic cholecystectomy.⁴ However, to our knowledge, no case has been reported so far that has described nerve blocks for non-cardiac surgery in a patient with EA.

Hypotension due to central neuraxial blocks (CNBs) can be hazardous because the routine treatment with fluids might lead to congestive heart failure, increasing the magnitude of right to the left shunt leading to hypoxemia. Ephedrine, a commonly used vasopressor, may precipitate supraventricular tachycardia in these patients (2). Hence, a pure alpha-agonist such as phenylephrine would be a better choice. Although general anesthesia (GA) is preferred

in severe cases, there is a possibility of supraventricular arrhythmias, hypotension during induction, and sudden pulmonary edema or cardiac arrest can be precipitated due to stress response to laryngoscopy and intubation. Hence, we have preferred the nerve blocks under ultrasound guidance to avoid significant hemodynamics implications. We avoided central venous catheterization as it has the risk of causing dysrhythmias and infective endocarditis, besides the technical difficulties. Also, we felt it was not necessary as we have avoided both GA and CNBs, which could cause hemodynamic instability. Bedside echocardiography is a proper cardiac monitor during surgery in such cases (3). In our case, we made it available in the operating room to assess the cardiac status if required.

Conclusion

The anesthetic technique and the selection of drugs and doses should be based on the patient's cardiac status. They should avoid a significant impact on hemodynamics when patients with cardiac anomalies such as EA undergo non-cardiac surgery. Ultrasound-guided regional blocks could be a better alternative than GA or CNBs wherever feasible in patients with EA.

Acknowledgment

None.

Conflicts of Interest

The authors declare that there are no conflicts of interest.

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