CONE PROCEDURE, A NEW APPROACH, A NEW HOPE FOR EBSTEIN ANOMALY PATIENT: A CASE REPORT

Jefferson Hidayat*, Andri Kurnia

Cardiothoracic Anesthesia, Department of Anesthesiology and Intensive Care, Faculty of Medicine, Universitas Indonesia, Jakarta, Indonesia

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Corresponding Author

Name	: Andri Kurnia
Email	: andri.dr@gmail.com
Mailing address	: Jl. Diponegoro No. 71
Telephone	: 081319458669
Fax	: -
ORCID ID	: 0000-0001-7820-4781

Abstract

Ebstein anomaly is a rare congenital disease affecting the cardiac structure involving the tricuspid valve, leading to right ventricular dysfunction. Patients with Ebstein's anomaly may have other structural abnormalities. Asymptomatic patients can be treated with conservative management, while symptomatic patients presenting with severe tricuspid regurgitation, NYHA class III-IV symptoms, and overwhelming tachyarrhythmias should undergo surgery. A 13-year-old girl presented with EA and ASD underwent a cone procedure with direct ASD closure and PFO creation. Cone procedure is a relatively new surgical approach in EA management and is preferred because it can be applied to various anatomical differences in EA. Managing the hemodynamic effects and anesthetic consideration during EA repair can be complex due to right ventricular dysfunction secondary to tricuspid regurgitation (TR). While opening the pericardium, the monitor showed VT, and the patient was given an internal defibrillation. Transesophageal echocardiography (TEE) was done intraoperatively to help evaluate ventricular function. The sternal closure was delayed due to cardiac edema. The procedure was done, but the patient's hemodynamic was unstable, so it was decided to delay the sternal closure. Many studies showed that the Cone procedure is a long-lasting repair of TR and creates significant functional improvement, resulting in low in-hospital mortality. TEE during the cone procedure has a vital role in evaluating the tricuspid valve's function after repair. Our patient's cone procedure was successfully done with reduced TR and good biventricular function, as shown in previous studies.

Keywords

Cone procedure, congenital heart disease, Ebstein anomaly, transesophageal echocardiography, tricuspid regurgitation

Introduction

Ebstein's anomaly (EA) is a rare congenital cardiac anomaly involving the tricuspid valve (TV), causing apical displacement of the tricuspid valve, leading to right ventricular dysfunction.[1] There is typically a Patent foramen ovale (PFO) or atrial septal defect (ASD) in patients with EA. Other structural cardiac abnormalities often seen in Ebstein's anomaly include pulmonary stenosis or atresia and ventricular septal defect.[2] Surgical intervention is recommended when there is evident enlargement of the right heart and a gradual decline in ventricular function.

Numerous surgical repair methods have been suggested due to the diverse range of anatomical variations found in cases of Ebstein's anomaly. Among the many approaches, Da Silva's Cone reconstruction stands out as it closely approaches anatomical restoration.[3] Cone procedure has shown better results in surgical outcome, as indicated by improved tricuspid regurgitation and reduced right ventricle size.[4] Another study has reported that the Cone procedure for Ebstein anomaly has low mortality and morbidity.[5] Transesophageal echocardiography (TEE) was performed in this case during the surgery to help evaluate ventricular function in line with the study by Sujatha et al., who stated that TEE has a critical role in formulating surgical plans, determining operability, and assessing tricuspid valve's function after repair.[6]

Case Report

A 13-year-old girl with Ebstein anomaly was admitted to undergo a Cone procedure. The patient had exertional dyspnea and cyanotic. The oxygen saturation was 85%, and the physical examination revealed a systolic murmur. The laboratory examination is within normal limits except for high hemoglobin and hematocrit levels. The chest radiograph showed cardiomegaly in all chambers [Figure 1], while The echocardiography showed EA appearance with typical atrialized right ventricle (RV), displacement septal RV leaflet toward apex, smallish RV cavity, small to moderate secondary atrial septal defect (ASD).

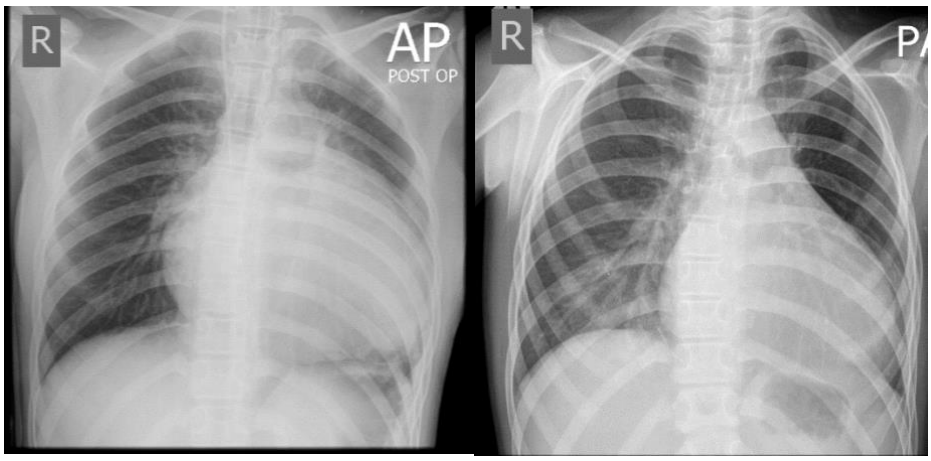

Figure 1. Left: CXR at op day right: fig at POD 14

The patient was admitted to the OR with basal hemodynamic of BP: 88/52 mmHg, HR: 95x/ min, SpO2: 82%. Co-induction was done with midazolam 1 mg and fentanyl 125 mcg. Still induction was done by sevoflurane 2%, and after the patient was unconscious, the muscle relaxant was given. After induction, an arterial line, a large bore IV, and a central line were inserted. Hemodynamic was (ABP): 98/66 (72) mmHg, HR: 96x/min, SpO2: 91 %, CVP: 3mmHg. The cone procedure is done with additional ASD closure. There is a septal annulus dilatation, and posterior tricuspid displacement into the RV was quite big. Secondary ASD 1.5 x 2 cm.

When the pericardium was opened, the monitor showed ventricular tachycardia (VT), and internal defibrillation 1x 20 Joule was given, converting the heart to Sinus rhythm, followed by supraventricular tachycardia (SVT) managed with lidocaine boluses. The cardiopulmonary bypass commenced, cross-clamp was initiated, and clear cardioplegia was given. The cone procedure was done to repair the tricuspid valve, and ASD was closed, leaving PFO. TEE was done after the procedure, showing no PFO shunt, mild-moderate TR with VC 0.32 cm, and PISA 0.38 (impaired RV contractility) [Figure 2].

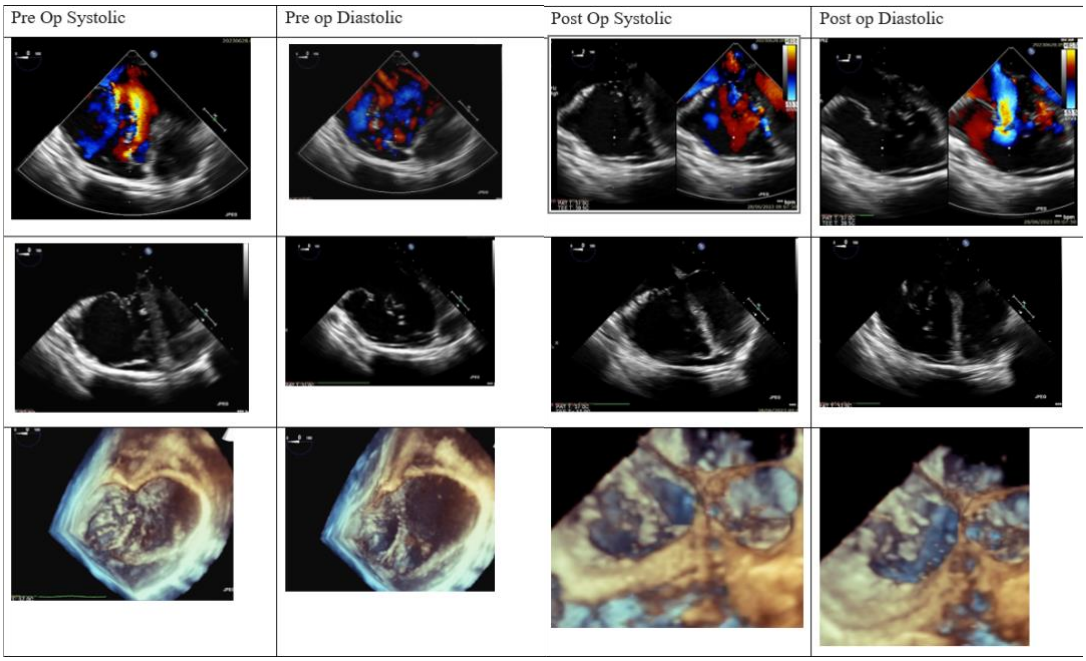

Figure 2. Transesophageal echocardiography before and after procedure

After the procedure, the patient was de-aired, cross-clamp off, and the CPB was weaned. The hemodynamic was HR: 126x/min, SpO2: 100%, ABP: 67/53 (45) mmHg, and

CVP: 2mmHg by adrenaline 0,2 mcg per kg per min and milrinone 0,2 mcg per kg per min. It was decided to delay the sternal closure due to cardiac edema and unstable hemodynamics. The patient's hemodynamic stabilized after two days of treatment, HR: 132x/min, SpO2: 100%, ABP: 97/62 (75) mmHg. The patient was discharged seven days post-surgery with good follow-up results.

Font Candara 12; Alignment justify; line spacing should be 1,15 throughout the text. Briefly summarizes why this case is unique and may include medical literature references.

Discussion

The patient had Ebstein anomaly, which is a rare congenital disease that involves malformations of the right ventricle and tricuspid valve that is characterized by multiple features that can exhibit an infinite spectrum of malformation. Abnormalities of the tricuspid valve and right ventricle include the following: (1) adherence of the tricuspid leaflets to the underlying myocardium (failure of delamination); (2) anterior and apical rotational displacement of the functional annulus (septal > posterior > anterior leaflet); (3) dilation of the "atrialized" portion of the right ventricle with variable degrees of hypertrophy and thinning of the wall; (4) redundancy, fenestrations, and tethering of the anterior leaflet; (5) dilation of the right atrioventricular junction (the true tricuspid annulus); and (6) variable ventricular myocardial dysfunction. These anatomical and functional abnormalities cause significant tricuspid regurgitation, which results in right atrial and right ventricular dilation and atrial and ventricular arrhythmias.[7]

As mentioned above, patients with EA can also have other cardiac defects like ASD or VSD, including PFO, which can cause right to left shunt. Asymptomatic patients may be managed medically with Follow-ups for years to come. Surgery was indicated in patients with symptomatic patients with New York Heart Association functional class III-IV, progressive deterioration of exercise capacity, congestive cardiac failure, cyanosis (systemic arterial oxygen saturation < 90%), paradoxical embolism in functional classes I and II with progressive cardiomegaly on chest X-ray (cardiothoracic ratio > 0.6), progressive right ventricular enlargement and onset or progression of uncontrolled tachyarrhythmias. Patients with Ebstein anomaly who have severe tricuspid regurgitation and atrial septal defects are recommended to undergo tricuspid valve surgery along with ASD closure.[8] As seen in this case, the patient has dyspnea, cyanosis, and cardiomegaly, so surgery was needed.

The cone procedure was selected as the surgery method. The cone reconstruction sets itself apart from other valvuloplasty methods due to its closest resemblance to natural anatomy disposition. The outcome of the cone reconstruction involves encircling the right atrioventricular junction with tricuspid leaflet tissue, amounting to 360° coverage,

allowing the leaflets to coapt properly, replicating the normal tricuspid valve anatomy. Additionally, the reconstructed tricuspid valve is reconnected at the true tricuspid valve annulus (atrioventricular junction), thus placing the valve's pivot point in its natural anatomical position. The thinned and translucent atrialized right ventricle is folded, eliminating any regions of abnormal movement. The excess right atrium is removed, restoring the right atrium's size to a more normal state. With minor exceptions of early postoperative right ventricular dilation, the cone reconstruction successfully reestablishes the visual appearance of the normal tricuspid valve structure.[9] The decision to opt for the cone procedure may depend on the Gose score value. Unfortunately, we cannot measure the Gose score as we cannot get the perfect view to count. There are other classifications, such as Carpentier, but looking at the patient's pre-op echo, we could conclude that the patient still has a chance for biventricular repair. If the atrialization of the right ventricle greatly disturbs the functionality of the right ventricle, sometimes less than the biventricle is selected. The procedure plan will also affect the anesthesia plan we selected.

Induction was done by still induction technique, by using sevoflurane inhalational anesthetic, co-inducted by midazolam and fentanyl. Midazolam is a sedative, causing retrograde amnesia, and also helps reduce the awareness and recall intra-anesthesia. Fentanyl was used as an analgetic, but the combination of midazolam and opioids confers a significant risk of vasodilatation, so we need to make sure the patient volume status is adequate. Other induction modalities, such as propofol and thiopental, could also be done, but with great vasodilatation and myocardial depression. Thus, an inhalational induction is a safe choice. Other agents, such as etomidate, trade the stable hemodynamic with adrenal cortical depression. Moreover, in Indonesia, etomidate is discontinued. For this reason, inhalational induction is still the technique of choice.

Pre-bypass, we faced right heart failure caused by mal-co-optation of the tricuspid valve. Therefore, it is necessary to improve right ventricle function simultaneously with decreasing its afterload.

The cone procedure has been shown to improve the right ventricular function, with residual mild-moderate TR might completely dissipate after follow-ups.[2,4] This procedure could also be considered for mildly symptomatic patients with significant valve regurgitation.[5,9] The challenge was maintaining the patient's hemodynamics; in Ebstein's anomaly, there is a ventricular malfunction due to prolonged tricuspid regurgitation. In our patient, the cone procedure was done, and ASD was closed, leaving a PFO for right-to-left shunt decompression of the right heart volume and pressure.

Post bypass, we also face the probability of right heart failure because of the tricuspid valve plication and right ventricle modification. Inotropes were needed to help the contractility; we used adrenaline up to 0.2 mcg per kg per minute. Also, we use milrinone 0.2 mcg per kg per min to help inotropy, lusitropy, and right heart afterload. Research also found that in chronic heart failure, there is downregulation of beta receptors, so the myocardium sensitivity to beta-agonists is lessened. At that point, milrinone also helps contractility by regulating the calcium influx and lusitropicity so the

ventricle can respond better to preload. The cone procedure helps the right ventricle to pump better than before, but the SIRS and right ventricle dysfunction must be coped.

The patient was unstable after the surgery with ABP 67/53 (45) mmHG, and it was decided to delay the sternal closure. Delayed sternal closure is a procedure to manage hemodynamic instability after executing complex heart surgery and has better outcomes than sternal reopening if monitored closely.[10] To improve ventricle function, we should also assess the preload and afterload of the cardiac function. So, it is essential to ensure adequate fluid and oxygen delivery. The uniqueness of cardiac surgery is that we can see the fluid adequacy directly, and if needed, we could test the loading response of the cardiac function at CPB weaning. Therefore, at the end of CPB, we could ensure adequate fluid and hemoglobin levels.

Open chest physiology was chosen because of the edema and hemodynamic instability in this case. The open chest lessens the lung compression to the edema heart, helping the heart's diastolic filling, waiting until the chest can be closed. However, this advantage is limited by the patient's discomfort, probability of patient-ventilator desynchrony, and risk of infection. That is why we need to do the closure as soon as the hemodynamic improves. After the chest was closed, the patient improved and was extubated on the third day of operation.

The Transesophageal echocardiography was done after the surgery, showing ventricular function improvement. TEE can help visualize the surgical plan and evaluate ventricular function after the procedure during the operation.[6] TEE can provide better visualization and information than Transthoracic echocardiography (TTE), which cannot be done in open chest surgery. A week after the surgery, follow-up echocardiography showed residual TR Mild with TVG 19 mmHg with good biventricular function.

This result aligns with other studies that showed that the Cone procedure has better prognosis outcomes.[5,9] Low morbidity and mortality were reported after the cone procedure.[9] The evaluation also shows the shrinkage of heart size, which shows that the cone procedure improved the heart pump function.

The cone reconstruction procedure is novel and has shown good morbidity and mortality results, as many studies support. Hemodynamic and anesthetic treatment should be monitored closely due to ventricular function abnormality. Transesophageal echocardiography helps to monitor and evaluate ventricular function after the valve's repair. Delayed sternal closure can help with hemodynamic instability after the procedure. Our patient has also successfully undergone the cone reconstruction procedure with good follow-up results.

Competing Interests

None declared.

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