

A Case of Cone Reconstruction and Aortic Valve Replacement for an Adult Patient Diagnosed with Ebstein's Anomaly Incidentally during Preoperative Examination of Severe Aortic Regurgitation – A Secondary Publication

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Abstract: Recently, there have been some reports that cone reconstruction can be performed in the repair of Ebstein's anomaly with acceptable results on a child. On an adult with Ebstein's anomaly, optimal surgical indication and choice of the operative procedure are controversial. A man in his seventies was diagnosed with Ebstein's anomaly incidentally during the preoperative examination of severe aortic regurgitation. We performed aortic valve replacement and cone reconstruction because his tricuspid regurgitation was moderate. There was no severe complication and he was discharged. No sign of recurrence has been observed after 4 months of follow-up. We present a case in which cone reconstruction and aortic valve replacement were successfully performed on an adult patient diagnosed with Ebstein's anomaly and severe aortic regurgitation.

Keywords: Ebstein's anomaly; Cone reconstruction; aortic valve replacement

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1. Case study

A 70-year-old male had a complaint of difficulty breathing and swelling of his lower legs. He has a family history of his father and brother suffering from stroke, and a history of neurologic syndrome, Sugamo segmental sclerosis, hypertension, and hyperlipidemia.

The patient was followed up for aortic regurgitation (AR) by his primary care physician but was referred to the Department of Cardiology because of gradually progressive dyspnea and worsening leg edema. The patient

was admitted to the hospital with a diagnosis of renal failure and heart failure. Ultrasonography revealed severe AR and moderate tricuspid regurgitation (TR). The cause of TR was found to be deviation of the septal apex toward the apex of the heart, and a diagnosis of Ebstein's disease was made. After induction of dialysis, the patient underwent surgery.

On admission, he was 158.6 cm tall, weighed 57.1 kg, NYHA grade I, SpO₂ 97% (room air), and had a clear breath sound on auscultation with a diastolic heart murmur of Levine 2/6 at the left border of the sternum. The abdomen was flat and soft with bilateral leg edema.

An electrocardiogram showed that the patient was in sinus rhythm with a heart rate of 61 beats/min, a PR interval of 224 ms, first-degree atrioventricular block, RV5 + SV1 of 5.03 mV, and left ventricular hypertrophy. There was no evidence of leg block or axial deviation.

Radiographs showed that the heart was enlarged with a cardiothoracic ratio of 67.2%, and the right first and second arches protruded (**Figure 1**). The diaphragmatic angles of the ribs were bilaterally acute, and there was no evidence of pulmonary congestion.



Figure 1. Preoperative chest X-ray with cardiothoracic ratio of 67.2%

The contrast-enhanced computed tomography (CT) showed that in addition to the calcification of the aortic valve and its surroundings, small and large calcifications were scattered throughout the aorta from the arch to the common iliac artery bifurcation. The diameter of the ascending aorta was 32 mm, and the peripheral aorta was within normal limits.

The transthoracic echocardiography (TTE) revealed a left ventricular ejection fraction (EF) of 58%, mild left ventricular wall thickening, and good wall motion. The aortic valve was calcified at the bicuspid apex, and moderate to severe AR was observed. TR was moderate, regurgitation velocity was 2.6 m/s (pressure gradient 26 mmHg), and tricuspid annulus diameter was 39.8 mm. The patient was diagnosed with Ebstein's disease and Carpentier type A. The Celermajorindex was 0.28, and the right ventricular wall motion was good (**Figure 2**). The mitral and pulmonary valves were abnormal. The mitral and pulmonary valves were normal. There was no interatrial traffic.

The patient had a severe AR with episodes of heart failure, and the decision was made to perform a large artery valvotomy. The patient was judged to be a candidate for surgery. The patient underwent surgery for the tricuspid valve. If the intraoperative findings showed mild plastering, he underwent valvuloplasty alone to control TR, otherwise, he underwent cone surgery.

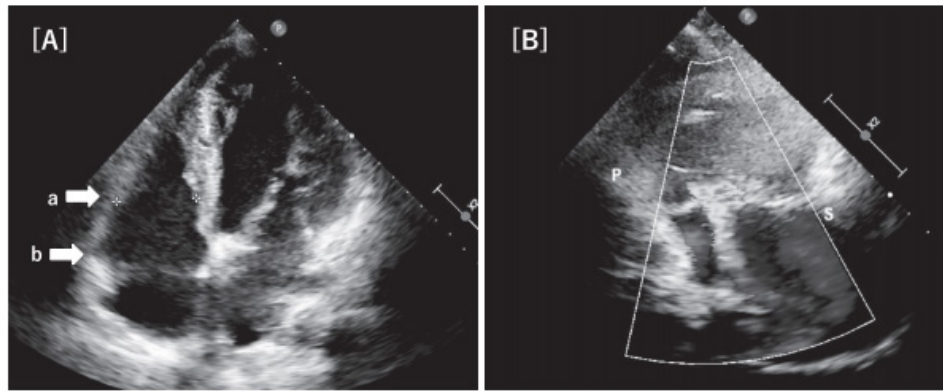


Figure 2.Preoperative transthoracic echocardiography. **(A)** In the four-chamber view, arrow a points to the functional tricuspid annulus, and arrow b points to the anatomical tricuspid annulus. Celermajor index = 0.28. **(B)** In the apical 2-chamber view, S is the septal leaflet, and P is the posterior leaflet. TR moderate.

1.1. Surgical intervention

The patient was placed in a supine position, ventilator-controlled, and under general anesthesia. The patient was approached through a median sternotomy, and extracorporeal circulation was established through the ascending aorta for blood supply and the superior and inferior vena cava for deblooding. A vent tube was inserted through the right superior pulmonary vein, and myocardial protection was provided by selective progressive and retrograde coronary perfusion.

Observation of the tricuspid valve after cardiac arrest reveals that the anterior apex is approximately 1/2 of the posterior apex. After cardiac arrest, the tricuspid valve was observed to be plastering extensively on the posterior half of the anterior apex, the posterior apex, and the septal apex. The anterior apex of the tricuspid valve was plastering over a wide area. The septal apex was not plastering. Because the plastering was not localized at the septal apex, regurgitation control was not possible by valve ring suture alone. The patient underwent Cone surgery as planned. The tricuspid valve ring was determined from its coloration, and the inner side of the ring was incised with a scalpel clockwise from the anterior to the posterior apex. The valve was detached from the right ventricular myocardium with a scalpel and shears, and all but the primary chordae were removed. Because the plastering right ventricular myocardium on the septal apex side and septal apex of the posterior apex was very thin and partially defective, the dissection was limited to about 2/3 of the posterior apex (**Figure 3**). The posterior apex was incised longitudinally from the valve ring to the apex, and the anterior septal commissure of the septal apex was incised longitudinally as well, and the severed ends were sutured with 5-0 monofilament thread to form a cone shape. Contralateral to the coronary sinus, the anatomic tricuspid valve ring was sutured with a 2-0 prejet polyester suture to the circumference of the cone, and the right atrialized right ventricle was subsequently longitudinally plicated. Because the valve ring was not histologically fragile, the Cone was pulled up over the sutured anatomic tricuspid valve ring and sutured directly and continuously with 5-0 monofilament thread. No regurgitation was observed during the water test. The aortic valve was tricuspid with minimal degeneration or calcification. The valve leaflets were resected and sutured with Inspiris RESILIA 23 mm (Edwards Lifesciences Irvine, California) using a 2-0 polyester suture with a single nodal suture. The patient was easily removed from the heart-lung machine. The operative time was 4 hours and 44 minutes, the ventilatory time was 3 hours and 6 minutes, and the aortic disconnection time was 2 hours and 27 minutes.

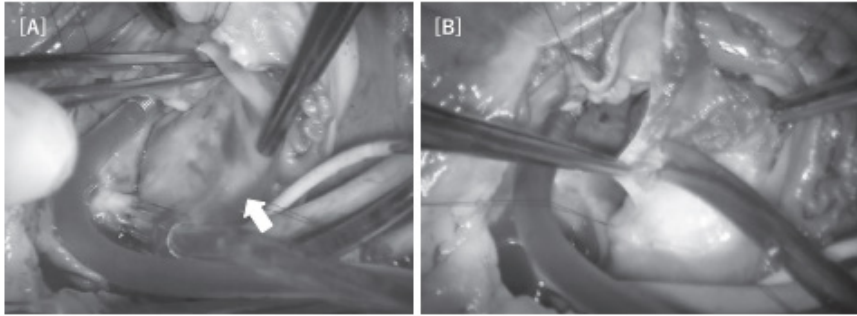


Figure 3. Intraoperative view. (A) The posterior leaflet was extensively plastered (arrow). (B) Anterior and posterior leaflets were detached. It was difficult to detach the whole posterior leaflet because of the thin right ventricular myocardium.

1.2. Postoperative course

The patient was intubated and sent to the intensive care unit. The patient was weaned from the ventilator on the first postoperative day. The patient was transferred to the general ward on postoperative day 2. Postoperative TTE on postoperative day 7 showed EF 55%, no cardiac depression, no AR, and mild TR. Immediately after surgery, the patient had occasional atrioventricular junction (AVJ)-like waveforms, but they gradually improved. The patient was discharged from the hospital on the 21st postoperative day. TTE was performed again on the 31st postoperative day, and TR was observed trivially to mildly along the septum between the anterior apex and septal apex, regurgitation velocity improved to 1.9 m/s (pressure gradient 14 mmHg), tricuspid valve ring diameter was 27.4 mm, and there was no obvious tricuspid stenosis (TS). The tricuspid valve ring diameter was 27.4 mm. Four months after surgery, the patient has been an outpatient without any significant changes (**Figure 4**).



Figure 4. Postoperative chest X-ray with a cardiothoracic ratio of 56.2%

2. Consideration

Ebstein's disease, first described by Wilhelm Ebstein in 1866, is an uncommon disease caused by abnormal development of the tricuspid valve and right ventricle with a frequency of less than 1% of congenital heart disease (1 in 200,000 births).

The disease is characterized by a lack of delamination of the tricuspid valve septal apex and posterior apex

in the right ventricular wall, a functional valve ring shifted toward the apex, right ventricular enlargement and thinning (right atrialization), anterior apex deformation, and right ventricular dysfunction due to narrowing, which may present in a variety of combinations and degrees ^[1,2]. According to Watson's analysis of 505 cases, 71% of patients aged 1–25 years and 60% of patients aged 25 years or older were in NYHA I-II with no major limitations in daily life ^[3]. In a study of 202 cases by Celermajor *et al.*, cyanosis and heart failure were the main symptoms in neonates and infants, while relatively mild symptoms such as heart murmurs and arrhythmias were more common in childhood and adulthood, and the annual mortality rate was 36% in the first year from birth to age 1, compared with 1.4% between 10 and 40 years of age ^[4]. The prognosis for Ebstein's disease, which does not require intervention until adulthood, is not so bad. Regarding the indications for surgery for Ebstein's disease in adulthood, Japanese guidelines for adult congenital heart disease and European guidelines recommend surgical intervention in patients with symptomatic or laboratory signs of right heart failure. In fact, the Mayo Clinic operated on 81 patients with Ebstein's disease aged 50 years or older between 1980 and 2010, of whom 85% were operated on in NYHA III-IV cases, and reported a relatively good 10-year survival rate of 71%.

In the report, the authors also suggested that early surgical intervention may be considered because of the poor prognosis in patients who underwent surgery at an advanced age or who did not show improvement in NYHA after surgery ^[5].

In the surgical treatment of simple Ebstein's disease after infancy, the cone procedure reported by da Silva *et al.* in 2007 has attracted much attention because, unlike the conventional technique of mono-cusp repair, this technique creates a new conical valve with complete dissection of almost all valve leaflets, which allows for diastolic central flow and better anatomy. This technique is considered to be a more anatomical repair, as it allows for central flow during diastole.

In the report, Cone surgery was performed in 40 patients, and the TR grade improved from 3.6 ± 0.5 to 1.2 ± 0.5 with no valve replacement at an average of 4 years of observation ^[6]. The choice between tricuspid valve repair or replacement in cases requiring surgical intervention in adulthood is still a matter of debate, but there are increasing reports of cone surgery being used as the surgical choice for tricuspid valve replacement ^[7,8]. Ozbek *et al.* compared cone surgery with other tricuspid valve procedures in adults with Ebstein's disease aged 20 years or older, with an average age of 44 years, and reported that cone surgery was superior in various aspects such as shutdown time, ICU stay time, hospital stay time, and TR survival, and cone surgery had no perioperative mortality ^[9]. Cone surgery was superior in various aspects, such as time of interception, ICU stay, length of hospital stay, and survival of TR.

In this case, Ebstein's disease was found incidentally during a close examination of the AR, and although the TR was moderate, we decided to intervene at the same time as AR surgery. Preoperative examination revealed relatively mild Ebstein's disease, and considering the patient's age and other factors, we assumed that TR could be controlled by valvuloplasty alone, but actual observation revealed extensive plastering. However, the plastering was extensive, and the intervention of the tricuspid valve in this case was very significant. The patient was treated with a cone procedure, which was a preferable alternative to a prosthetic valve because the patient was on hemodialysis. However, in this case, the patient was prepared for bioprosthetic valve replacement, but if the available valve leaflets are limited or TS is suspected during the intraoperative TEE evaluation, switching to tricuspid valve replacement should be considered.

Regarding surgical intervention for simple TR associated with left heart surgery other than tricuspid valve surgery, aggressive simultaneous surgery may be considered even if the patient has moderate TR at risk ^[10]. As mentioned above, Ebstein's disease can develop without any particular symptoms, but in Japan, with its aging population, cases such as the present case are rare but still possible. In the adult population with mostly

degenerative cardiac disease, the addition of Ebstein's disease with its many variations may make the treatment strategy more complex and difficult. This case suggests that even in asymptomatic Ebstein's disease, surgical intervention should be considered when other left ventricular procedures are necessary, depending on the severity of the disease.

3. Conclusion

Aortic valve replacement and cone surgery were performed simultaneously for Ebstein's disease, which was found incidentally during a thorough examination of patients with severe aortic regurgitation, with good results.

Disclosure statement

The authors declare no conflict of interest.

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