Cardiology in the Young

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Transcatheter pulmonary valve implantation in tetralogy of Fallot and Ebstein's anomaly with one and a half ventricular repair

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Brief Report

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Abstract

The coexistence of tetralogy of Fallot and Ebstein's anomaly is extremely rare. There are only a few case reports in the literature, and surgical options for the treatment are controversial. There is insufficient data on long-term follow-up of patients and management of complications. In this case report, we present a 20-year-old adult with operated tetralogy of Fallot, Ebstein's anomaly, and Glenn anastomosis who underwent transcatheter pulmonary valve implantation for severe pulmonary insufficiency.

The coexistence of tetralogy of Fallot and Ebstein's anomaly is extremely rare.[1] There are only a few case reports in the literature, and management of this condition is controversial. There is insufficient data on long-term results of patients and management of complications. In this case report, we present a 20-year-old adult treated with a transannular patch repair, patch closure of the ventricular septal defect, and Glenn anastomosis when he was 2 years old with diagnosis of tetralogy of Fallot and Ebstein's anomaly, who underwent transcatheter pulmonary valve implantation for severe pulmonary insufficiency and right ventricular dysfunction. To the best of our knowledge, this is the first report of transcatheter pulmonary valve implantation in a patient with operated tetralogy of Fallot, Ebstein's anomaly, and Glenn anastomosis.

Case

A 20-year-old male patient with operated tetralogy of Fallot and Ebstein's anomaly was presented with exertional dyspnea. He had undergone surgery with a transannular patch repair, patch closure of the ventricular septal defect, and Glenn anastomosis when he was 2 years old. Physical examination revealed a 3/6 to-and-fro murmur on the left upper sternal margin and pectus excavatum. The patient's heart rate was 98/minute, blood pressure was 110/56 mmHg in the right arm, and O_2 saturation was 98%. Electrocardiography and a 24-hour Holter examination revealed a first-degree atrioventricular block and complete right bundle branch block (QRS was 140 msec) and normal sinus rhythm with rare supraventricular premature beats. Echocardiography (Philips Epic ultrasound system, Philips Healthcare, Inc., Andover, MA) demonstrated tricuspid valve dysplasia with low implantation of posterior leaflet, enlarged right heart cavities, and slightly decreased right ventricular systolic function (Fig 1a, video 1). Colour Doppler showed moderate tricuspid regurgitation, severe pulmonary insufficiency, and stenosis in the proximal right pulmonary artery (Fig 1b and c, video1). Cardiac magnetic resonance imaging detected that the right heart cavities were dilated than normal (right ventricular end-diastolic volume: 205 ml/m^2, right ventricular end-systolic volume: 115 ml/m^2), a decreased right ventricular ejection fraction (38%), significant pulmonary regurgitation (regurgitation fraction 62%), and an aneurysm of right ventricular outflow tract.

The patient was evaluated for transcatheter pulmonary valve implantation and underwent cardiac catheterisation via the right femoral vein and right jugular vein access. Right ventricle and pulmonary artery angiograms showed dilated right ventricular outflow tract, crossing pulmonary arteries, severe pulmonary insufficiency, and proximal right pulmonary artery stenosis (Fig 1d, video 2). Right ventricular outflow tract sizing was performed by 30 × 40 mm TyShak baloon (NuMED, Hopkinton, New York). After the balloon was inflated to nominal pressure, balloon interrogation and contrast injection were performed through the long sheath (Fig 1e, video 2). The main pulmonary artery was measured 29 mm at the narrowest part. Haemodynamic data are presented in Table 1. Our case was evaluated in terms of transcatheter intervention to the proximal right pulmonary artery and transcatheter pulmonary valve implantation in the first angiography. It was observed that the right pulmonary artery pressure was lower than the left due to the stenosis in the proximal right pulmonary artery and the Glenn shunt was functional. In this circumstance, transcatheter pulmonary valve implantation was planned without interfering with the patient's proximal right pulmonary artery.

Table 1. Haemodynamic data during the first cardiac catheterisation

	Pressures (mmHg)
Right ventricle	30/(0–2)
Right pulmonary artery	17/4 (9)
Left pulmonary artery	19/12 (14)
Aorta	83/54 (64)
Left atrium (mean)	5
Right atrium (mean)	5

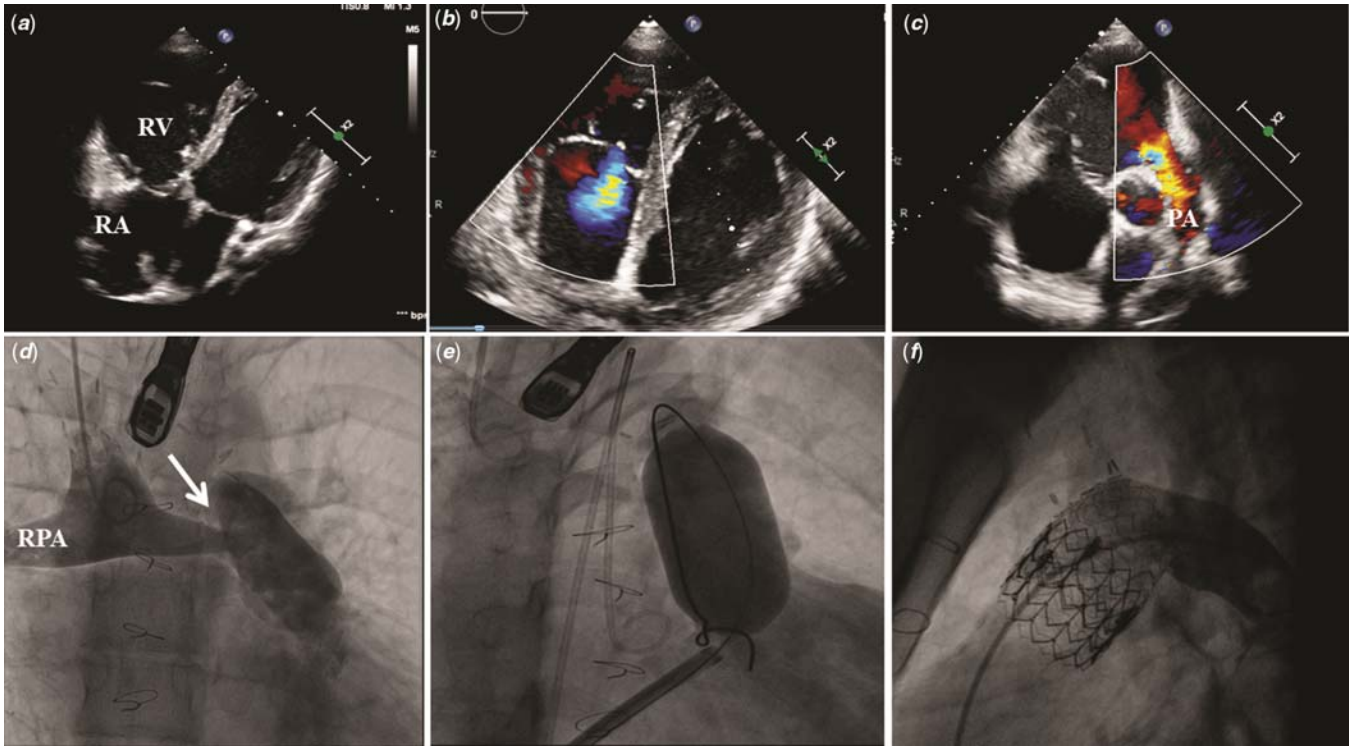

Figure 1. (***a***) Echocardiography demonstrates enlarged right heart cavities and Ebstein's anomaly. (***b***) Colour Doppler shows moderate tricuspid regurgitation. (***c***) Echocardiography demonstrates severe pulmonary insufficiency. (***d***) Pulmonary artery angiogram through the Glenn anastomosis in the anteroposterior projection shows severe pulmonary insufficiency and proximal right pulmonary artery stenosis (arrow). (***e***) Contrast injection after the balloon was inflated to nominal pressure. (***f***) Pulmonary artery angiogram in the lateral projection after valve deployment shows valve position and no residual pulmonary regurgitation; *RA: right atrium, RV: right ventricle, MPA: main pulmonary artery*.

The second cardiac catheterisation was performed under general anaesthesia via a right femoral vein approach. A 6 F right Judkins catheter (Cordis Corporation, Miami, FL) was advanced into the distal part of the left pulmonary artery and a Backup-Meier guidewire (Boston Scientific Corp., Natick, MA) was placed inside the pulmonary artery. An 18F long sheat was inserted to the femoral vein and a 48-mm Andra XXL stent (Andramed, Reutlingen, DE) was mounted on a 30 × 45 mm Z-med-2 balloon (B. Braun Medical Inc.) and implanted at the distal portion of the main pulmonary artery. A 29-mm Edwards SAPIEN XT valve (Edwards Lifesciences, Irvine, CA, USA) (balloon was filled with 2 ml of additional volume) was deployed to the stent. An angiography demonstrated a competent pulmonary valve without regurgitation (Fig 1f, video 2). The total fluoroscopy time was 16.4 minutes. The patient was monitored overnight. His echocardiogram showed normal pulmonary valve haemodynamic and the patient was discharged home on day 2. The patient's fatigue regressed and O_2 saturation remained the same after 1 year of follow-up.

Discussion

Until today, there have only been a few case reports about patients with tetralogy of Fallot and Ebstein's anomaly in the literature and no consensus on surgical treatment was reported.[2-4] Chan and colleagues published the first surgical correction in a 7-month-old girl with tetralogy of Fallot in the presence of Ebstein's anomaly.[1] On the other hand, there have also been no publications about long-term results and management of complications in patients with operated tetralogy of Fallot and Ebstein's anomaly. In this case report, we presented the first case of transcatheter pulmonary valve

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implantation in a patient with operated tetralogy of Fallot, Ebstein's anomaly and one and a half ventricular repair.

There may be several treatment options in our patient with operated tetralogy of Fallot and one and a half ventricular repair. The first decision to be made is between surgical and transcatheter options and, we preferred transcatheter pulmonary valve implantation for our patient. In the second stage, there were two alternative access points for valve implantation: one jugular vein due to Glenn anastomosis or femoral vein. After the first angiograms, it was concluded that the jugular vein approach was not suitable because of its complex pathway and insufficient distance for loading the valve over the 20 Fr Nova-flex delivery system. However, in patients with tetralogy of Fallot and one and a half ventricular repair, it should be kept in mind that if the enlarged right atrium prevents the long sheet from advancing, the procedure could be performed through Glenn anastomosis.

Another problem noticed after the first angiogram was the stenosis of proximal right pulmonary artery. After discussing the patient's case in the paediatric cardiology consul, it was concluded that the right pulmonary artery would be filled by the Glenn anastomosis and the antegrade blood supply through the left pulmonary artery would be sufficient.

In conclusion, transcatheter pulmonary valve implantation has recently become an alternative method to surgery in patients who have undergone transannular patch repair or conduit repalcement. As in our case, transcatheter pulmonary valve implantation can be applied safely and effectively in selected cases with one and a half ventricle repair.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951120004436

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Conflicts of interest. None.

Ethical standards. The authors assert that this work complies with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008. This case was approved by the patient's family.

References

1. Chan KC, Walker DR, Gibbs JL. Surgical correction of tetralogy of Fallot in the presence of Ebstein's anomaly of the tricuspid valve. Int J Cardiol 1989; 25: 242–243.
2. Abid A, Dhiab M, Fehri W. [A rare association: tetralogy of Fallot and Ebstein's anomaly]. Arch Mal Coeur Vaiss 2006; 99: 77–79.
3. Lai CH, Wu JM, Yang YJ. Ebstein's anomaly of the tricuspid valve in combination of tetralogy of Fallot: total correction in infancy. Ann Thorac Surg 2007; 83: 304–306.
4. Wei LY, Shih JC, Wu ET, Chen SJ, Chen YS, Huang SC. Surgical Correction for a Neonate With Ebstein's Anomaly Associated With Tetralogy of Fallot. Ann Thorac Surg 2017; 104: e79–e81.