Bidirectional cavopulmonary shunt as a rescue procedure for right ventricular endomyocardial fibrosis

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1. Case report

A 31-year-old male patient was referred to us from a local clinic with a 7-year-history of NYHA class III dyspnea for further investigation and treatment. He had no past history of rheumatic fever during childhood or cyanosis at birth. Physical examination showed his jugular veins were not engorged. Low intensity of S1 sound was heard and S2 sound was widely split without fixation. A soft pansystolic murmur was heard at the right lower sternal border. No abnormal breathing sounds were heard in either lung field. Shifting dullness accompanied by hepatomegaly and mild pitting edema was also detected. Cardiorespiratory ratio was 0.5 on the plain film and no other specific findings were noted. ECG showed sinus rhythm with a pattern of right bundle branch block in the precordial lead. Echocardiographic findings indicated that the septal leaflet of the tricuspid valve (TV) was apically displaced downward, creating an impressively large right atrium (RA) and very small right ventricle (RV) (Fig. 1). The motion of the interventricular septum (IVS) was paradoxical and grade II/IV tricuspid regurgitation (TR) was observed. Pulmonary hypertension was not found. Cardiac catheterization showed leftward displacement of the TV and very small RV chamber. RA pressure was 20/10 mmHg and RV systolic pressure 32 mmHg, with an end-diastolic pressure of 10 mmHg. A large amount of TR flow and infundibular stenosis were noted in each cardiac cycle.

Based on the above, especially echocardiographic and cardiac catheteric evidence, we suspected the present case to have Ebstein’s anomaly and decided on surgical treatment. Intraoperative findings were consistent with endomyocardial fibrosis (EMF) rather than Ebstein’s anomaly. In the RV cavity, yellowish thick endocardial fibrotic membrane covered all of the RV chamber, and extended to the posterior leaflet of TV and the base of the pulmonary valve. The annulus was dilated with poor leaflet coaptation. The anterior leaflet of TV was thickened and destroyed. The posterior and septal leaflets had downward displacement due to the traction by fibrotic membranous tissue, which seemed to hinder RV contraction. However, there was no notable morphology, indicative of Ebstein’s anomaly, such as typical atrialized RV created by downward displacement of the hinge point of the septal and posterior leaflet. The annulus was located in the normal position.
Surgery was performed under standard cardiopulmonary bypass (CPB) with moderate hypothermia and cold blood antegrade cardioplegia to protect the myocardium. Through small vertical ventriculotomy and tricuspid valvulotomy, fibrotic endocardial tissue was resected and leaflets were mobilized. The tricuspid valve was not repaired since displaced posterior leaflet was closely attached to the posterior wall of the RV and the anterior leaflet was too distorted to be reconstructed. Accordingly, the tricuspid valve was replaced by a prosthesis. With no notable intraoperative events, the present case exhibited a too poor hemodynamic profile from the cardiopulmonary bypass, with systolic blood pressure decreased to 50 mmHg and central venous pressure increased to 17 mmHg. The right ventricle was severely dilated and feeble, due it was assumed to poor contractility of the RV. We immediately performed bi-directional cardiopulmonary shunt (BCPS) to reduce the overload in the right ventricle. Since then, the patient has been weaned from cardiopulmonary bypass with no notable hemodynamics.

The present case was extubated on postoperative day 1 and discharged on postoperative day 13 with no sign of RV failure, and showed no further episodes of RV failure during an 8-month follow-up period. The diagnosis was established as endomyocardial fibrosis through surgical and pathologic evaluation of resected material (Fig. 2).

2. Discussion

Since EMF was first reported by Davies in Uganda in 1948, it has been known to be prevalent in tropical countries. As an acquired disorder, EMF is characterized by endocardial fibrosis as well as myocardial infiltration. Moreover, EMF is frequently involved in partial obliteration of one or both ventricular cavities [1].

EMF has an etiology different from Ebstein’s anomaly, although its clinical and hemodynamic profiles are very similar to those of Ebstein’s anomaly in cases where EMF involves RV only [2]. In Korea, though extremely rare, EMF is preoperatively misdiagnosed as Ebstein’s anomaly based on a similar hemodynamic profile.

Many studies have reported operative procedure as well as early and intermediate results of surgical treatment for EMF [3–5]. These studies have established endocardial decortication and atrioventricular valve replacement or repair as the procedure of choice for surgical treatment, although they did not include bi-directional or total cavopulmonary shunt. Amit et al. [6] reported a patient with EMF who underwent bi-directional Glenn shunt with an 8-year follow-up result, and Kumar et al. [7] performed total cavopulmonary connection for a case of EMF. However, they did not perform intracardiac procedures.

BCPS diverts 30–40% of the systemic venous return from the right ventricle [8] and thereby decreases the RV work index. Since BCPS gives rise to elevation of pulmonary artery flow, it is routinely performed to diminish RV preload in patients with RV hypoplasia or dysfunction. It is nowadays well established that BCPS decreases intraoperative mortality and the rate of revision surgery in high-risk patients with Ebstein’s anomaly [9]. The hemodynamic profile of the present case was similar to that of Ebstein’s anomaly. Therefore, we concomitantly performed BCPS to overcome acute right heart failure after endocardectomy and tricuspid valve replacement, with an excellent outcome. This produced a decreased right atrial pressure.
from 16 to 8 mmHg with no further elevation of superior vena cava pressure.

To date, the long-term outcome and the indications for BCPS have not yet been established, although the present case showed that BCPS was effective for saving the life of a patient with low cardiac output and acute RV failure.

References


Appendix A. ICVTS on-line discussion

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Message: The Authors should be congratulated for the brilliant idea, used not only to acutely wean the patient from cardiopulmonary bypass, but also to provide him with a good long term prognosis. In fact the Authors were very prompt to apply to an unusual situation the surgical technique of the “one-and-half ventricular repair”, more and more frequently used for complex congenital heart defects. The medium-term results, whenever available, seem to be quite encouraging on the tolerance of the hemodynamics imposed by the “one-and-half ventricular repair”, and this makes this approach suitable also as a life-saving procedure in all situations with acute right ventricular dysfunction at the end of cardiopulmonary bypass, or even some hours or days after surgery. Of course this approach is only possible in the presence of normal pulmonary artery pressure and resistance. It would be interesting to know if the Authors have any data available from the follow-up, particularly regarding the right ventricular function. It would be interesting to know if the Authors have any data available from the follow-up, particularly regarding the right ventricular function. It is possible that, weeks or months far from the operation, there is some degree of recovery of the right ventricular function. If this is the case, it would be interesting for the readers to know if the improved right ventricular function, with better antegrade pulsatile flow in the pulmonary artery, in any way interferes with the functioning of the bidirectional cavopulmonary shunt.