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Case Report

Hemi-Fontan or Bidirectional Cavopulmonary Shunt for Right Ventricular Failure after Mitral Valve Replacement and Acute Ascending Aortic Dissection: Report of Two Cases

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Abstract

Right ventricular failure due to right coronary artery disease, right ventricular hypertrophy, stunning, abnormal septal motion, myocardial infarction, or non-homogeneous distribution of cardioplegia is an uncommon but serious complication of open heart surgery. We report a patient with severe right ventricular hypertrophy secondary to severe mitral valve stenosis and another patient with detachment of the right coronary artery due to the dissection of the ascending aorta. The patients developed right ventricular failure, which persisted after surgery and rendered weaning from cardiopulmonary bypass unsuccessful. Through a hemi-Fontan, or bidirectional cavopulmonary shunt, and an intra-aortic balloon pump, the patients were successfully weaned from cardiopulmonary bypass. This shunt may be an alternative to a right ventricular assist device in some patients with right ventricular failure. The long-term outcome and the indication of bi-directional cavopulmonary shunt has not been confirmed, although it is believed to be effective for saving the life of patients with low cardiac output and acute right ventricular failure. In our cases, six months following the operation, there was some degree of recovery of the right ventricular function. In long-term follow-up, however, it would be interesting for the authors 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.

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Introduction

The bidirectional cavopulmonary shunt, or the hemi-Fontan, was first described clinically by Hopkins for tricuspid atresia in 1985.[1] Since then, this method has been used, albeit infrequently, as a palliative treatment for Ebstein's anomaly or severe right ventricular (RV) failure in adult patients provided that there is no pulmonary artery hypertension.[2, 3] The hemi-Fontan can augment pulmonary artery flow without increasing RV work. This shunt has one important advantage in that it requires no anticoagulation post-cardiotomy, when there is prolonged pump time and diffuse coagulopathy,[4] as was the case in the two patients introduced herein. We performed a bidirectional cavopulmonary shunt,

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or hemi-Fontan, to wean 2 patients from cardiopulmonary bypass (CPB). The first case suffered from RV dysfunction after mitral valve replacement and the second case was a patient with ascending aorta dissection and detachment of the right coronary artery, complicated by acute inferior myocardial infarction.

The First Case Report

A 42-year-old woman with a past medical history of moderate mitral stenosis was admitted to our hospital with respiratory distress. A left atrial clot was diagnosed, and the patient underwent urgent surgery for clot extraction and mitral valve replacement. During anesthesia induction, the patient became hypotensive and required extra inotropic drug support. Her central venous pressure (CVP) was 15 mmHg.

The chest was opened through a median sternotomy. RV function appeared moderately impaired, and there was severe RV hypertrophy and no pulmonary hypertension. CPB was established using the ascending aorta and bicaval cannulation. During hypothermia, the aorta was cross-clamped and cardioplegia was established antegradely and retrogradely through the aortic root and right atrium. The mitral valve, which was rheumatically stenotic, was resected via the left atrium while preserving the posterior papillary muscles. The valve was thereafter replaced with a 31-mm Carbomedix mechanical prosthesis.

With no notable intraoperative events, the patient exhibited too poor a hemodynamic profile to be weaned from CPB: her systolic blood pressure dropped to 50 mmHg, CVP rose to 30 mmHg, and RV became severely dilated and feeble. An intra-aortic balloon pump (IABP) was inserted to minimize left atrial pressure. Despite preserved left ventricular (LV) function, it was impossible to conclude CPB due to persistent RV failure. At 70% of the CPB flow, the patient's CVP increased to 23 mmHg with normal pulmonary artery pressure and high left atrial pressure (>10 mmHg). Under these circumstances, CPB was resumed and mechanical causes (paravalvular leak) were ruled out by transesophageal echocardiography (TEE), and other correctable causes such as gas exchange problems, electrolyte and acid-base disturbance, and arrhythmia were corrected. Additionally, echocardiography was performed before the use of the IABP in the patient and disclosed RV dilatation, reduced systolic motion of the RV free wall, and systolic movement of the interventricular septum to the right. Mean pulmonary artery pressure was normal. Be that as it may, the patient's systemic output did not improve with high-dose inotropic drug use.

With diffuse bleeding and normal pulmonary artery pressure, the patient appeared to be a good candidate for the hemi-Fontan.[5] The hemi-Fontan was performed between the superior vena cava and the right pulmonary artery, and the inotropic drug administration as well as the IABP was continued.

It became possible to wean the patient CPB after 185 minutes. After the procedure, RV contractility, as determined by inspection, was improved and the dosage of the inotropic drug was reduced to 10 μg/kg/min with a CVP of 22 mmHg.

The postoperative course was without major complications except for the occurrences of atrial fibrillation, which were converted with the aid of the external defibrillator. The patient's CVP decreased to 15 mmHg by the second postoperative day. The IABP was removed on the second postoperative day, and she was extubated on postoperative day 4 with a CVP of 12 mmHg. TEE during the postoperative period showed reduced RV function by day 7, after which time contractility improved slowly to an ejection fraction of 30% on day of 15.

The patient was discharged on the 20th postoperative day with Warfarin. Three months after discharge, she underwent heart catheterization, which demonstrated a patent hemi-Fontan shunt. Hemodynamic examination disclosed a pulmonary artery pressure of 30/15 mmHg.

The Second Case Report

A 66-year-old man with a past history of uncontrolled hypertension was admitted to another hospital with acute chest pain. Acute inferior myocardial infarction was diagnosed and treated with heparin. An electrocardiogram showed ST-segment elevation in leads II and III as well as aVF, which subsequently changed to an inverted T wave with a Q wave. One week later, severe chest pain and increasing dyspnea prompted transthoracic echocardiography, which revealed acute ascending aorta dissection. The patient was then transferred to our hospital, and he underwent emergency surgeryin the operating room.

Soon after intubation, the patient experienced severe hypotension with a CVP of 16 mmHg, for which he required extra vasopressor support. A rapid median sternotomy was performed under CPB. Ante grade cold blood cardioplegia was instituted to arrest and protect the heart and systemic hypothermia to 28 °C was achieved. RV function appeared markedly impaired, and there was moderate bloody pericardial effusion. During systemic cooling, the ascending aorta was cross-clamped and cardioplegia was given directly into the coronary ostia. The aortic valve was found tricuspid, and the right coronary artery (RCA) ostium was detached by a false lumen. The Bental operation was performed and the ostium of the RCA was reconstructed and anastomosed to the composite graft. Coronary circulation was resumed after a cross-clamp time of 76 minutes. The patient was re-warmed and an attempt was made to wean him off CPB. This proved unsuccessful as the heart was barely able to maintain a systolic blood pressure of 60 mmHg, CVP of 20 mmHg, and pulmonary artery pressure of 20 mmHg, despite adequate preload and high inotropic and IABP support. After

sufficient reperfusion and time so as to allow contractility to resume, RV contractility was still impaired.

Using a segment of the great saphenous vein, a coronary artery bypass graft (CABG) was made to the RCA. Nevertheless, it was impossible to conclude CPB due to persistent RV failure despite preserved LV function. The patient's systemic output did not improve with high doses of adrenaline, Dobutamine, and Milrinone. Hemodynamic ally, the RV failure was reflected by right atrial pressure > 20 mmHg, left atrial pressure < 10 mmHg, cardiac index < 1.8 L/min/m^2, and decreasing cardiac output, developing in the setting of high pulmonary arterial pressure and CVP. Tricuspid regurgitation might have contributed to the RV insufficiency. Intraoperatively, the RV appeared distended, a finding that was correlated with the echocardiographic evidence of the interventricular septum extending into the LV.

With a prolonged CPB time, coagulopathy, diffuse bleeding, and normal pulmonary artery pressure as well as the absence of an RV assist device (RVAD), the patient seemed to be a good candidate for the hemi-Fontan. A bidirectional cavopulmonary shunt was performed between the superior venacava and the right pulmonary artery, and the administration of the vasopressors as well as the IABP use was continued.

It became possible to wean the patient from CPB after 167 minutes. Immediately after the procedure, RV contractility, as determined by inspection and TEE, was improved. The dosage of the inotropic drugs was reduced to minimum, and the CVP decreased to 13 mmHg. The postoperative course proved eventful because of acute renal failure. The IABP was removed on the 4th postoperative day, and the patient was extubated on postoperative day 7 with a CVP of 16 mmHg. Repeat TEE studies during the postoperative period showed almost severe RV contractility dysfunction. Doppler sonography of the renal artery revealed obstruction of both renal arteries due to dissection. Contractility then improved slowly to an estimated RV ejection fraction of40% on day 12. Despite peritoneal dialysis, the patient expired due to complications of renal failure.

Discussion

The rise in the incidence of predisposing factors such as LV failure, CPB use, myocardial infarction, and ischemic condition has led to an increase inthe number of patients with decompensated RV failure. Mitral valve replacement frequently involves the mitral valve apparatus, as is the case in papillary muscle resection and changes in the LV geometry, and this influences successful weaning from CPB. RV failure is a possible consequence of post-cardiotomy stunning, pulmonary hypertension, RV hypertrophy, emboli, myocardial infarction, geometric changes after mitral valve replacement, or catheter-induced coronary sinus rupture.[6-10]

In the first case, we reported a female patient with RV failure due to RV hypertrophy and stunning, who underwent mitral valve replacement. In the second case, there was detachment of the coronary artery due to dissection. RV dysfunction unresponsive to medical therapy occurs in approximately 10-15% of patients with acute dissection submitted for surgery. It may be also observed in patients with myocardial infarction and post-cardiotomy cardiogenic shock and Aprotinin usage.[9, 10] In addition to the impairment of the circulatory function by the inadequate delivery of systemic venous return to the pulmonary circulation, RV failure is responsible for ventricular overloading, which represents an important limitation for better recovery of the myocardialfunction.[10] Coronary artery detachment occurs in 10% of acute ascending aorta dissections. If surgery is performed expeditiously, by restoration of the coronary flow blood, the myocardium will be preserved. On the contrary, when the myocardium is damaged irreversibly due to delay in surgery, RV failure will ensue. In our 2 patients, difficulty in CPB weaning, in spite of high-dose inotropic drug and IABP use, prompted us to undertake a hemi-Fontan.

RV failure is not uncommon after heart surgery: it occurs in 1% of all cardiac surgeries.[11] When the myocardium is damaged irreversibly due to myocardial infarction or reversibly due to post-cardiotomy stunning or non-homogenous distribution of cardioplegia, RV failure may occur.[1, 5] LV unloading with the IABP or a left ventricular assist device (LVAD) can also precipitate RV failure.[12] Post-cardiotomy RV failure is seen less frequently than LV failure, and it is important to differentiate between reduced contractility and increased after load in its treatment. The IABP is useful in the treatment of mild to moderate cases of RV failure because the IABP reduces left atrial pressure and thus pulmonary pressure.[13] Buck berg and collogues14 reported that postoperative septal dysfunction was the underlying reason for RV failure after cardiac surgery and demonstrated that this adverse event impaired the twisting function essential for RV ejection against increased vascular resistance.

The septum constitutes approximately 40% of the mass of the heart, and myocardial stunning is the cause of its dysfunction in the absence of an excitation-contraction abnormality.[14] O'Neill and associates[15] reported that the RVAD might be the most effective solution in this situation. Parr and colleagues[16] utilized the RVAD in a patient with LV aneurysm, who developed RV failure after open heart surgery.

The literature lacks a consensus on the choice between surgery and medical therapy for the treatment of RV failure. The observation of improved survival of patients with pulmonary hypertension associated with a foramen ovale has led to this hypothesis that atrial septostomy, which decompress the RV and increases right-to-left shunting, may

be helpful in severe RV failure.[17] We would not recommend this method for RV failure because response to this method in RV failure is variable; consequently, atrial septostomy should be deemed palliative. Another method for the treatment of severe RV failure is heart transplantation. Patients with refractory RV failure associated with left heart failure or patients with arrhythmogenic RV dysplasia and refractory tachyarrhythmia in the absence of severe pulmonary hypertension may be considered for heart transplantation.[18] Bypassing the RV by venting the right atrium and infusing blood into the pulmonary artery is another treatment method for RV failure. Nevertheless, this method has the potential risk of overwhelming pulmonary edema, prolonged ventilator dependency, pneumonia, and prolonged mechanical ventilation.[19] The use of the IABP in the pulmonary circulation to unload a failing RV has recently stimulated clinical interest; we, however, had no experience with this method and there is evidence that the IABP is not effective in treating right heart failure.[20] RV dysfunction significantly increases mortality following mitral valve replacement and remains a major cause of acute mortality after cardiac transplantation and LVAD placement. RV failure is a progressive disorder that starts with myocardial injury, stunning, myocardial infarction, inappropriate distribution of cardioplegia, neurohormonal activation, cytokine activation, complement activation, and ventricular remodeling, all of which may contribute to the exacerbation of the injury.[21]

Until the ongoing research has ushered a better understanding of the pathophysiological basis of this syndrome, the hemi-Fontan is a palliative surgery. The use of the hemi-Fontan in the treatment of severe RV dysfunction has been reported in multiple studies by Tagakaki et al.[22] and Danton et al.[23] The Hemi-Fontan, or the bidirectional cavopulmonary shunt, seems to be capable of adequately maintaining the pulmonary blood flow in the presence of RV failure; it also seems to reduce LV dysfunction by limiting RV dilation and restoring interventricular septal position and LV cavity shape, as was suggested by Tagakaki and Denton.[22, 23] In this regard, the decrease in RV pressure overloading observed with the hemi-Fontan anastomosis is encouraging and confers a suitable perspective forthe use of this surgical approach as a bridge to myocardial function recovery. Application of the hemi-Fontan reduces RV volume and significantly improves LV systolic impairment observed with RV infarction. Zehenderet al.[24] emphasized the importance of RV dilation and ventricular septal contraction in the low cardiac output after RV ischemia. The mechanism whereby the hemi-Fontan effectively augments LV contractility is likely to be a reduction in RV volume via resumption in interventricular septal motion and position and the LV cavity shape. The successful outcomes of the Fontan operation, in which adequate pulmonary blood flowcan be achieved by way of passive systemic venous pressure without RV contribution, have lent further support to this hypothesis. However, there is now increasing evidence that RV functionis an important determining factor in the outcome of many cardiac conditions. Zehender et al.[24] showed that mortality after acute myocardial infarction was significantly increased with RV involvement, with 26% versus 64% complications without and with RV involvement, respectively.

The postoperative course of our first patient was complicated by atrial fibrillation. Despite medical intervention, continuous atrial fibrillation resulted in hemodynamic disturbances, which were subsequently resolved with the external defibrillator. Postoperative atrial fibrillation is a common arrhythmia and in this settlements related to heart failure, pulmonary edema, atelectasis, and hypoxia,[25] RV function remains poorly characterized. Furthermore, the mechanisms whereby RV failure contributes to hemodynamic insult have yet to be clearlyidentified.[26-28]

Conclusion

In conclusion, the removal of RV overload via the RV exclusion procedure could confer effective volume loading, restore a normal geometrical shape, and improve the systolic function of the LV; it is, therefore, expected to augment the systemic output. The physiological restoration of the LV size, geometry, and function in our study population hints at the promise which this procedure holds as a useful therapeutic option for isolated end-stage RV failure caused by multiple factors in post-cardiotomy RV dysfunction.

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