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CLINICAL REPORT

Anesthetic management of pulmonary valve replacement for pulmonary regurgitation in six patients with surgically repaired tetralogy of Fallot

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Abstract Tetralogy of Fallot (TOF) is the most common form of cyanotic congenital heart disease (CHD) encountered in CHD patients surviving into adulthood. A number of patients with surgically repaired TOF have significant pulmonary regurgitation (PR) that can lead to right ventricular (RV) dilatation, RV failure, and arrhythmia. We describe the anesthetic management for pulmonary valve replacement (PVR) in six PR patients with surgically repaired TOF. Although all patients had dilated RVs and depression of RV ejection fraction preoperatively, and arrhythmia and unexpected bleeding perioperatively, they could tolerate a well-managed PVR operation. Anesthesiologists should be aware of the multiplicity of comorbidities, sequelae, and residua in patients with surgically repaired TOF. RV function should be monitored using transesophageal echocardiography, and inotropic vasodilators and alpha-adrenergic agents should be administered, as appropriate. Arrhythmias, vascular injury during removal of adhesions during re-sternotomy, and bleeding from collateral vessels are also frequent complications.

Keywords Tetralogy of Fallot · Adult · Right-sided heart failure · Pulmonary valve · Pulmonary regurgitation

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Introduction

The population of adult congenital heart disease (ACHD) patients requiring cardiac surgeries is increasing; hence, the anesthetic management of patients with ACHD has become a major topic of discussion in this decade [1–5]. Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease, which also has the most favorable prognosis [6]. Some patients with surgically repaired TOF require a second cardiac operation in adulthood. Surgical repair of TOF includes right ventricular (RV) outflow tract reconstruction and closure of the ventricular septal defect (VSD). Despite the surgery, however, the cardiac condition of many patients progresses, with significant pulmonary regurgitation (PR) that can lead to RV dilatation and dysfunction, arrhythmias, and sudden cardiac death. PR is the most common indication for late reoperation [7]. Information about the anesthetic management of such patients is inadequate. We introduce a case series of PVR for PR patients with surgically repaired TOF.

Case series

After approval by the Ethics Committee of our hospital, the hospital database was evaluated to retrospectively identify six patients with surgically repaired TOF, double outlet RV-type TOF, and pulmonary atresia with VSD who underwent PVR from 2011 to 2013 (Table 1). We reviewed their most recently recorded data from echocardiograms, cine cardiac magnetic resonance images (CMR), or cardiac catheterization before surgery. The patients' demographic data are presented as mean ± standard deviation or median value (range). Preoperatively, all the patients had greater than moderate PR and mild TR. RV end-diastolic volume index

Table 1 Demographics data and medical history of patients

No.	Age (years)	Sex	BMI (kg/m^2)	Initial diagnosis	Initial TOF repair	Age at repair (years)	Interval between repair and PVR (years)	Further cardiac surgeries	Other cardiac diseases	Other medical history
1	42	F	22	TOF	TAP	8	34	Embolization of residual BT shunt at 13 years, residual VSD closure and TVP at 23 years, stenting of PA at 42 years	CA of AFltr	Left recurrent nerve paralysis
2	19	F	17	DORV/PS	TAP	3	16	Embolization of collateral circulation for left PA occlusion at 19 years		
3	58	M	21	DORV/PS	TAP	20	38	Residual VSD closure at 20 years	Nonsustained VT, AFltr	Diabetes mellitus, liver cirrhosis
4	20	M	15	PA/IVS	TAP	4	16	Balloon dilation of PA at 15 years	CA for AT	
5	28	F	20	TOF	TAP	4	24	Stenting of PA at 28 years	Pacemaker for AV block	Epilepsia, asthma
6	58	M	16	TOF	TAP	25	33		2° AV block	

M male, *F* female, *TOF* tetralogy of Fallot, *ASD* atrial septal defect, *DORV* double outlet right ventricle, *PS* pulmonary stenosis, *PA/IVS* pulmonary atresia with intact ventricular septal, *TAP* transannular patch, *PVR* pulmonary valve replacement, *BT* Blalock–Taussig, *VSD* ventricular septal defect, *TVP* tricuspid valve plasty, *PA* pulmonary artery, *CA* cryoablation, *VT* ventricular tachycardia, *AFltr* atrial flutter, *AT* atrial tachycardia, *AV* atrioventricular

(RVEDVI) was significantly increased to 193 ± 34 ml/m^2 (normal value, 108 ml/m^2), and RV ejection fraction (EF) was decreased to 34 ± 9 % (normal value, >50 %). Brain natriuretic peptide (BNP) was increased to 61 ± 39 pg/ml (normal value, <18.4 pg/ml).

In the operating room, all patients were monitored for ECG, pulse oximetry, arterial blood pressure, central venous pressure (CVP), and transesophageal echocardiography (TEE). Midazolam was used as the induction agent in five patients (4 ± 0.7 mg), and one patient was anesthetized with propofol (50 mg). Anesthesia was maintained with fentanyl (70 ± 21 μg/kg), remifentanil (0.1–0.5 μg/kg/min), and propofol, with adjustment of the bispectral index. Neuromuscular relaxation was provided using rocuronium. Inotropic vasodilators (dobutamine or milrinone), alpha-adrenergic agents (noradrenaline or phenylephrine), and coronary vasodilators (nicorandil or nitroglycerin) were administered to all patients. With the aim to reduce pulmonary vascular resistance, the intraoperative ventilatory strategy included a high oxygen concentration, avoidance of hypercapnia, and optimum lung volume and pressure.

All patients underwent concomitant procedures together with the PVR (Table 2), and the femoral vessels were exposed before re-sternotomy to enable commencement of emergent cardiopulmonary support (CPS) for accidental cardiovascular injury. Emergent CPS was required in one patient. Anesthesia time was 1,009 (range, 540–1,097) min, surgical time was 884 (397–964) min, and cardiopulmonary bypass time was 397 (141–512) min. In three patients, the aorta was clamped for 231 (212–291) min. Blood loss was 2,788 (543–8,881) g, and the amounts of transfusion of red cells, fresh frozen plasma, and platelet concentrations were 2,160 (240–3,120), 1,440 (320–3,360), and 500 (200–800) ml, respectively. The perioperative complications are summarized in Table 2.

Despite invasive surgery, during the ICU period none of the patients had hemodynamic instability requiring assisted circulation treatment, renal failure requiring continuous hemodiafiltration, or microbiologically diagnosed systemic inflammatory response syndrome.

Duration of ICU stay was 5 (3–18) days, and mechanical ventilatory support was required for 3 (2–16) days. Postoperative echocardiograms did not show PR in any of the patients, and they were discharged within 60 days after their operation. Left ventricular EF did not significantly alter between before (56 ± 8 %) and after (62 ± 1 %) the operation by echocardiography.

In four patients, RVEF and RVEDVI were assessed after surgery (period after surgery, 194 ± 29 days). The parallel for RVEF between before and after surgery showed no significant differences; however, RVEDVI was significantly reduced from 195 to 123 ml/m^2 ($p = 0.0247$).

Table 2 Operation and complications

No.	Operation	Exposed vessels before re-sternotomy	Intraoperative complications	Postoperative complications
1	PVR, TVP, CA	FA	None	Tracheotomy for glottic edema
2	PVR, TVP, MVP	FA, RSCA	3° AV block	3° AV block
3	PVR, TVP, CA	FA	None	AFltr terminated with electric defibrillator
4	PVR, TAP, CA, asc-Ao replacement	FA	Damage to innominate vein and ascending aorta	None
5	PVR, CA, PM upgrade	FA	None	None
6	PVR, aortic root and arch replacement	FA, RSCA	None	Hemothorax caused by bleeding from collateral vessels, nonsustained Af

PVR pulmonary valve replacement, *TVR/TVP* tricuspid valve replacement/tricuspid valve plasty, *CA* cryoablation, *MVP* mitral valve plasty, *asc-Ao* ascending aorta, *PM* pacemaker, *FA* femoral artery, *RSCA* right subclavian artery, *AV block* atrioventricular block, *AFltr* atrial flutter, *Af* atrial fibrillation

Discussion

Preoperative assessment of right heart failure

The key pre-PVR cardiac issues in patients with surgically repaired TOF are shown in Table 3 [1]. In particular, right heart failure is the most important issue complicating the anesthetic management of these patients.

Our data clearly indicated the presence of right heart failure in all the patients, manifesting as high RVEDVI, low RVEF, and high BNP levels. The mechanism of this dysfunction can be explained by the surgical history. Previously, the initial TOF repair tended toward aggressive right ventricular outlet tract enlargement, despite the resultant severe PR. TR may be secondary to RV dilation from the PR [7]. Reportedly, plasma BNP levels were significantly higher in patients with TOF than in controls [8]. The timing of PVR was often delayed after taking into account the need for subsequent replacement of the bioprosthetic valve every 10 years [2]. The optimal timing of PVR in significant PR patients remains uncertain [7]. Cheung et al. [9] reported that surgical PVR in patients with repaired TOF had been associated with significant decrease in RVEDV but no changes in the RVEF. This study reached the same result.

Table 3 Key cardiac issues before pulmonary valve replacement (PVR)

Cardiovascular events	Complications
Supraventricular events	Atrial flutter
	Atrial fibrillation
	AV block
	Tricuspid regurgitation
Ventricular events	RV dilation and dysfunction
	Residual RVOT obstruction
	Residual VSD
	Sustained VT
Pulmonary artery	Stenosis or hypoplasia
Others	Progressive AR
	Dilated ascending aorta
	Sudden cardiac death

RV right ventricle, *RVOT* right ventricular outflow tract, *VSD* ventricular septal defect, *VT* ventricular tachycardia, *AR* aortic regurgitation

RV failure is commonly diagnosed by the detection of typical patterns of CVP derangement in the context of low cardiac output or systemic hypotension [10]. TEE is of particular importance in this setting. Excessive loading causes deviation of both the interatrial septum and the interventricular septum to the left, creates significant TR, and enlarges the right atrium (RA) and RV. In our case series, RV function was evaluated by TEE observation of the volume change in the RA and RV during systole and diastole and visualization of blood flow. Inotropic support is usually administered to treat the development of RV failure. Milrinone and dobutamine are cardiotonics that are known to increase myocardial contractility, cause vasodilation, and decrease pulmonary vascular resistance, and hence should be used in this setting. The vasodilating effect of milrinone may facilitate LV filling and improve cardiac output, although these agents sometimes cause systemic hypotension. Alpha-adrenergic agents, such as phenylephrine or norepinephrine, are also used to maintain adequate arterial and coronary perfusion pressure. We used milrinone (0.2–0.5 μg/kg/min), dobutamine (0.5–7 μg/kg/min), norepinephrine (0.02–0.04 μg/kg/min), and bolus phenylephrine (0.05–0.1 mg) for circulatory support, and monitored cardiac function by TEE and CVP. With this management, none of our patients developed worsening of RV failure, PR, or TR.

Re-sternotomy

Re-sternotomy and removal of adhesions around the heart and aorta increase the risk of catastrophic hemorrhage [11]. In such cases, the femoral artery and vein should be exteriorized before re-sternotomy to enable emergency CPS. In this report, emergent CPS was required in one patient soon after injury to the innominate vein during synechiotomy. In the same patient, the ascending aorta was also damaged, and therefore ascending aorta replacement was performed as a concomitant procedure. Reportedly, re-sternotomy for pediatric cardiac surgery resulted in cardiac laceration in 10 of 192 patients (5.2 %) [12]. Patients with surgically repaired TOF have an increased risk of arrhythmia, especially during synechiotomy. Hence, we recommend that defibrillator electrode pads be preemptively applied on every patient. In this report, none of the patients required intraoperative defibrillation.

Postoperative complications

Arrhythmias are the most frequent postoperative complication. One patient in our case series experienced transient atrial flutter, another patient experienced transient atrial fibrillation, and another patient experienced complete atrioventricular block. Bédard et al. reported that atrial tachycardia and heart block are frequent in patients with repaired TOF [13]. One patient had hemothorax caused by bleeding from collateral vessels that required embolization. This patient had the initial repair at age 25; hence, long-term cyanosis probably led to the development of a collateral circulation [13]. Thus, attention must be paid to the possibility of bleeding from collateral vessels in adult ACHD patients.

In conclusion, anesthesiologists should be aware of the multiplicity of comorbidities, sequelae, and residua in patients with surgically repaired TOF. Although patients usually have a dilated RV and depression of RVEF, they can tolerate a well-managed PVR operation. Anesthesiologists can prevent intraoperative worsening of right heart failure by careful TEE monitoring and appropriate administration of inotropic vasodilators and alpha-adrenergic agents. With this approach, none of our patients experienced worsening of RV failure, although arrhythmias, vascular injury during removal of adhesions, and bleeding from collateral vessels were unavoidable complications.

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