Case report - Congenital

Cardiac resynchronization therapy for management of congestive heart failure after repair of tetralogy of Fallot in an elderly patient

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Abstract

A 65-year-old woman underwent total correction of tetralogy of Fallot with closure of ventricular septal defect, right ventricular outflow patch, pulmonary valve replacement and tricuspid valve repair. Pacemaker implantation using epicardial electrodes was simultaneously needed because she had complete atrioventricular block. The postoperative course was excellent. Eighteen months postoperatively, she was admitted with severe congestive heart failure and frequent ventricular arrhythmia. Echocardiography and cardiac catheterization revealed depressed left ventricular function caused by conduction delay. Cardiac resynchronization therapy with a defibrillation system was effective for improvement of left ventricular function. Ventricular contractility rapidly recovered to normal, and the patient has been asymptomatic for two years since implantation.

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1. Introduction

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease (CHD) in childhood. Complete surgical repair can be performed in adult patients to improve symptoms and survival. However, postoperative cardiac dysfunction, ventricular arrhythmia and sudden death are sometimes observed over the long-term. Management of postoperative arrhythmias and maintenance of ventricular function are important to achieve preferable long-term outcome.

We present herein the case of an elderly TOF patient with postoperative heart failure who was successfully managed by cardiac resynchronization therapy with a defibrillation system (CRT-D).

2. Case report

A 65-year-old woman was admitted to our hospital complaining of dyspnea and transient loss of consciousness. She had been diagnosed with TOF in her 50s. Electrocardiography (ECG) on admission revealed complete atrioventricular block with right bundle branch block. Her heart rate was 43 beats/min. A temporary percutaneous pacemaker was inserted immediately. Cardiac catheterization revealed a systolic right ventricular pressure of 99 mmHg, a right ventricular outflow tract (RVOT) systolic pressure gradient of 54 mmHg, pulmonary capillary wedge pressure of 5 mmHg and pulmonary arterial pressure of 15/9/11 mmHg (systolic/diastolic/mean). PaO₂ was 47.9 mmHg under nasal administration of oxygen at 3 l/min. The patient had well-developed confluent pulmonary arteries. Left ventricular contraction was normal. General condition of the patient was improved with administration of intravenous diuretic agents and temporary pacing. Considering the risk of thrombosis and infectious endocarditis due to the temporary pacing electrode in the malformed heart, we decided to perform surgery.

The patient underwent total correction of TOF with closure of the ventricular septal defect, reconstruction of RVOT with transannular equine patch and PVR using a stented bovine xenograft. The tricuspid valve was repaired concomitantly using an annuloplasty ring. An atrio-ventricular dual chamber permanent pacemaker was implanted using epicardial electrodes on both the right atrial and right ventricular free walls.

Postoperative course was good, and the patient was discharged from our hospital on postoperative day 13 without any complications. Echocardiography on discharge revealed no residual shunt, no tricuspid regurgitation, and no RVOT stenosis. Left ventricular ejection fraction (EF) was 54.4% and left ventricular end-diastolic volume index (EDVI) was 62.5 ml/m².

Eighteen months postoperatively, she was re-hospitalized with severe congestive heart failure despite adequate oral intake of digitals and diuretics. ECG showed a long QRS duration of 200 ms and a normal heart rate of 80 beats/min in DDD mode (Fig. 1a). Chest radiography demonstrated pulmonary congestion. Premature ventricular contraction was frequently identified. Cardiac catheterization showed...
unexpected depression of left ventricular function with EF of 15.3% and EDVI of 214.4 ml/m² (Fig. 2a). As coronary angiography and valvular function of the aortic and mitral valves were normal, we considered that asynchrony between the two ventricles might interfere with ventricular function. We therefore decided to provide CRT-D to improve left ventricular function and prevent fatal ventricular arrhythmia. The LV electrode was positioned in the posterolateral vein via the left subclavian vein. RA and RV leads were inserted from the left subclavian vein into the right auricle and right ventricular apex, respectively. The CRT-D was programmed to DDD mode with lower rate of 70 beats/min, paced and sensed AV intervals of 130 ms and 100 ms, and VV interval of 0 ms. After CRT-D implantation, symptoms of congestive heart failure disappeared and EF improved to 32.2% on echocardiography. ECG showed improved QRS duration to 134 ms (Fig. 1b).

The patient has since remained asymptomatic as of two years after implantation of CRT-D. Follow-up cardiac catheterization revealed that EF and EDVI had improved to 58.1% and 91.5 ml/m², respectively (Fig. 2b).

3. Discussion

The optimum age for surgical correction of TOF has been suggested as three to six months [1], although some patients with TOF survive to late adulthood without surgical intervention [2]. Surgical repair of TOF in elderly patients is frequently associated with arrhythmic events postoperatively and strong impacts on early and late mortality [2, 3].
Management of ventricular dysfunction and fatal arrhythmia is an important issue during follow-up for TOF patients. Two important management strategies, electrophysiological and pharmacological approaches, have been advocated for this condition. Restoration of pulmonary valve function by PVR is beneficial for both right ventricular dysfunction and refractory arrhythmia after the repair of TOF [4, 5]. Although the efficacy of PVR has been confirmed in younger patients, this approach may also be advantageous for elderly patients with TOF.

Biventricular synchronization using CRT was recently introduced and its efficacy in the management of congestive heart failure has been proven in large clinical trials [6, 7]. The concept of CRT is characterized by synchronized contraction of two ventricles, correction of left ventricular conduction delays and coordinated contraction of the two ventricles. Epicardial pacing on the hypertrophied right ventricle might induce severe conduction delay in the left ventricle. We deduced that the conduction delay caused reversible left ventricular remodeling in this patient. As the CRT-D also works as a defibrillator, mortality also appears to be reduced in comparison with single CRT [6]. CRT can be performed in pediatric and CHD patients, and has been recently shown its possible beneficial effects for those patients [8–10]. However, as anatomical and electrophysiological features of CHD vary enormously in patients, the indication of CRT-D for CHD patients is still unknown. Gatzoulis et al. showed in a multicenter study that a risk factor for ventricular tachycardia after repair of TOF was QRS duration >180 ms. Factors associated with sudden cardiac death were higher age at repair and longer QRS duration [3]. They also reported QRS rate of change during follow-up as an additional predictor of ventricular tachycardia and sudden death. Considering these risk factors as indications for CRT-D, we believe CRT-D implantation is a preferable option in the management of left ventricular dysfunction with conduction delay for TOF patients after surgical repair.

In conclusion, complete surgical repair for an elderly TOF patient was performed safely. Maintenance of pulmonary valve function by PVR and biventricular synchronization using CRT-D implantation with endocardial electrodes was useful to improve long-term ventricular function.

References