

Standard cardiac resynchronization therapy with a second right ventricular lead for severe right ventricular heart failure in 2 patients with repaired tetralogy of Fallot



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Introduction

Although early surgical repair for tetralogy of Fallot (rTOF) has significantly improved clinical outcomes and long-term survival,¹ progressive right ventricle (RV) dysfunction driving morbidity and mortality is often observed in adults with rTOF.² Management of RV heart failure is occasionally refractory to even appropriate medical therapies.³ RV mechanical dyssynchrony that is commonly observed in rTOF patients with right bundle branch block (RBBB) promotes depressed contractility and remodeling, and it is proposed as a desirable therapeutic target.⁴ We present 2 cases of rTOF complicated by severe RV heart failure, which were effectively managed with standard cardiac resynchronization therapy (CRT) with a second RV lead (CRT-RV).

Case presentations

Case 1

A 66-year-old man was admitted to the hospital because of severe ascites and hypotension, which were uncontrollable despite high doses of diuretic medication and catecholamine infusion. He had undergone rTOF combined with aortic valve replacement at the age of 38. A 12-lead

electrocardiogram (ECG) during sinus rhythm showed complete RBBB and prolonged PQ interval of 250 milliseconds (Figure 1A). A transthoracic echocardiogram revealed preserved left ventricular (LV) size and an ejection fraction of 63% with paradoxical septal motion, but a moderate or severe impairment of RV contractile function (Table 1), and marked conduction delay of the RV free wall compared with that of the septal wall, consistent with mechanical dyssynchrony in the RV (Figure 1B). The activation map of the RV during sinus rhythm made by using a CARTO XP 3-dimensional electroanatomic mapping system (Biosense Webster, Diamond Bar, CA) also demonstrated the conduction delay of the RV basal lateral wall compared with that of the septal wall, consistent with electrical dyssynchrony in the RV (Figures 1C and 1D). Right heart catheterization revealed an RV pressure of 42/17/31 mm Hg (systolic/diastolic/end-diastolic) with a dip and plateau pattern, a mean right atrial (RA) pressure of 26 mm Hg, and a pulmonary capillary wedge pressure of 20 mm Hg.

To perform CRT to specifically improve RV dyssynchrony, we successfully implanted 2 bipolar leads (5076 CapSureFix Novus) in the RV apex and basal lateral wall by using a manually shaped stylet, and a 4194 Attain OTW bipolar lead in the anterolateral coronary sinus. We then connected the distal poles of each RV basal lateral and LV lead as cathode and anode, respectively, using a 5866-3811 Y-adapter to the LV port, and the tip of the RV apical lead to the RV port of a Consulta CRT-P C3TR01 device (Medtronic, Minneapolis, MN) (Figure 1E and 1F). CRT was achieved by atrial synchronous, simultaneous biventricular multifocal pacing with sensed atrioventricular delay of 100 milliseconds, resulting in a shortening of the QRS width (Figure 1G)

After the CRT-RV, RV heart failure was improved, resulting in discontinuation of catecholamine 1 week later (Table 1). An echocardiogram showed normalization of RV dyssynchrony (Figure 1H) and improvement of RV contractile function (Table 1). The patient has remained

KEYWORDS Cardiac resynchronization therapy; Dyssynchrony; Echocardiography; Electroanatomic mapping; Right ventricular heart failure; Repaired tetralogy of Fallot

ABBREVIATIONS %DFT = diastolic filling time; BNP = B-type natriuretic peptide; CRT = cardiac resynchronization therapy; CRT-RV = standard cardiac resynchronization therapy with a second right ventricular lead; CTR = cardiothoracic ratio; ECG = electrocardiogram; LV = left ventricle; NYHA = New York Heart Association; RA = right atrium; RBBB = right bundle branch block; rTOF = surgical repair for tetralogy of Fallot; RV = right ventricle; RVFW = right ventricular free wall; SEP = apical septal wall; TAPSE = tricuspid annular plane systolic excursion; TRPG = tricuspid regurgitant pressure gradient (Heart Rhythm Case Reports 2016;2:76–79)

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KEY TEACHING POINTS

- Right ventricular (RV) mechanical dyssynchrony is proposed as a desirable therapeutic target for surgical repair of tetralogy of Fallot (rTOF) patients who have right bundle branch block or RV apical pacing complicated with RV heart failure. This condition can be visualized using transthoracic echocardiography or electroanatomic mapping, both of which reveal marked delay in the conduction of the RV free wall versus that of the septal wall.
- Right-sided cardiac resynchronization therapy (CRT) by application of additional pacing at the RV free wall is effective for the treatment of RV heart failure that is refractory to medical therapy, in patients with rTOF with RV dyssynchrony.
- To perform right-sided CRT using standard CRT equipment with only 3 ports, one can connect the RV apical lead to the RV port to stabilize ventricular sensing and connect the other 2 leads at the RV free wall and the LV-to-LV port via the Y-connector.

asymptomatic for 8 months after the implantation. Compared with massive ascites before the CRT-RV, obviously

decreased ascites showed in the results of follow-up computed tomography (Figures 1I and 1J).

Case 2

A 56-year-old woman had been repeatedly hospitalized because of exacerbations of leg edema for several years. She had undergone rTOF at the age of 25, followed by closure of a residual ventricular septal defect and tricuspid annuloplasty for tricuspid regurgitation at the age of 48. A dual-chamber pacemaker was also implanted because of atrial fibrillation with a slow ventricular response. On the patient's admission to the hospital, a 12-lead ECG showed RV paced rhythm with atrial fibrillation (Figure 2A). A transthoracic echocardiogram revealed preserved LV size but mildly reduced LV contraction with an ejection fraction of 50% and paradoxical septal motion, as well as mild impairment of RV systolic function. The analysis of longitudinal strain with 2-dimensional speckle tracking demonstrated RV dyssynchrony (Figure 2B).

CRT was performed with a system similar to that used in case 1, except for the use of a previously implanted 1452T RV lead (St Jude Medical, St. Paul, MN) (Figures 2C and 2D). CRT was achieved by atrial asynchronous, simultaneous biventricular multifocal pacing, resulting in a shortening of QRS width (Figure 2E). After CRT-RV, RV heart failure was improved, and there has been no rehospitalization

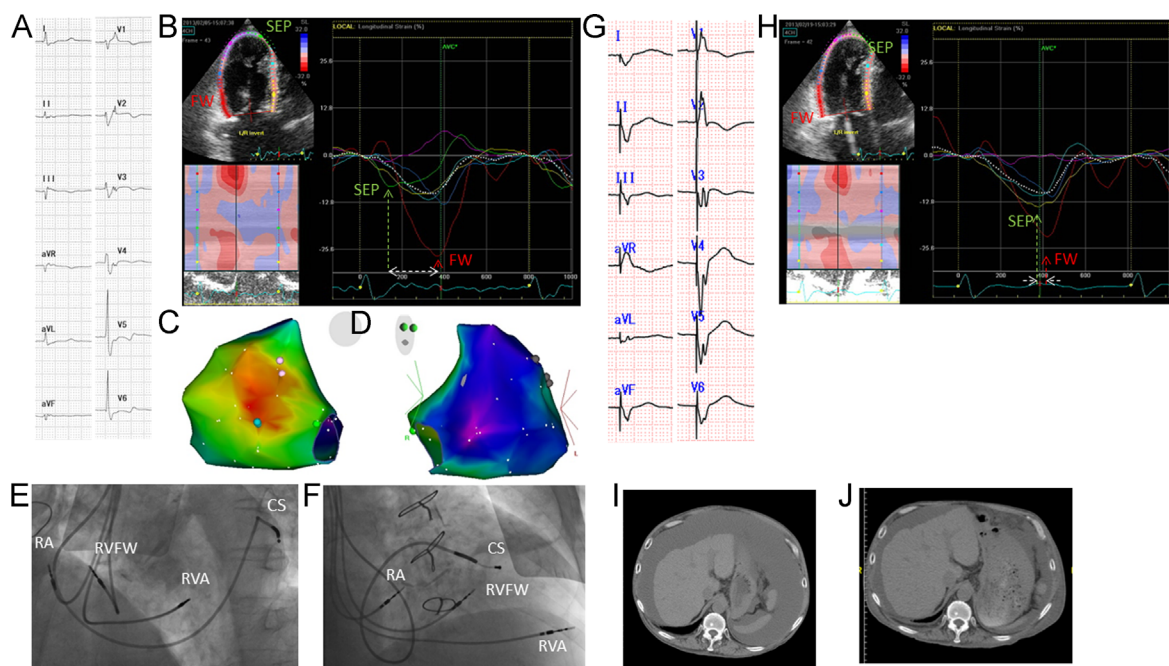


Figure 1 Data in case 1. **A and G:** 12-lead electrocardiograms before and after the cardiac resynchronization therapy (CRT), respectively. The QRS duration of 164 milliseconds before the implantation shortened to 140 milliseconds during CRT. **B and H:** 2-dimensional speckle tracking–derived displacement curves of right ventricle (RV) and septal segments in an apical 4-chamber view before and after the implantation, respectively. Positive and negative values indicate the location of each sample relative to baseline, inside and outside the RV cavity. Red and green dotted arrows indicate negative peak of the curves of RV free wall (RVFW) and apical septal wall (SEP), respectively. Contraction delay of RVFW relative to the SEP (indicated by horizontal bidirectional arrows) was 228 milliseconds, whereas the delay was improved by 72 milliseconds after CRT. **C and D:** Activation maps of the RV septum and RVFW, respectively, during sinus rhythm with right bundle branch block (RBBB) in a right anterior oblique view. Left-to-right transseptal wavefront breakthrough from the RV midseptum, with propagation to the basal lateral wall with a delay of 129 milliseconds, compared with that of the RV septal wall. **E and F:** Fluoroscopic views in left and right oblique projections showing lead positions. **I and J:** Abdominal computed tomography before and after the implantation, respectively. CS = coronary sinus, RA = right atrium, RVA = RV apex.

Table 1 Clinical parameters and echocardiographic measurements of right atrium and right ventricle before and after standard cardiac resynchronization therapy with a second right ventricular lead

	Case 1		Case 2	
	Before CRT	After CRT	Before CRT	After CRT
NYHA functional class	IV	II	III	II
Body weight, kg	60.4	55.0	61.0	56.5
Systolic blood pressure, mm Hg	76	109	135	115
CTR, %	58	52	76	72
BNP, pg/mL	131.7	111.2	148.3	86.2
RA dimension, mm	63	57	69	70
RV dimension (midcavity), mm	46	43	48	51
RV function				
Systolic pressure, mm Hg	43	32	50	39
TRPG, mm Hg	28	17	35	31
Fractional area change, %	17.6	27.6	45	48
TAPSE, mm	8.3	12.3	14	12
S', cm/s	6.1	5.5	8.1	5.8
%DFT	58	68	NM	NM

BNP = B-type natriuretic peptide; CTR = cardiothoracic ratio; NYHA = New York Heart Association; RA = right atrium; RV = right ventricle; TAPSE = tricuspid annular plane systolic excursion; TRPG = tricuspid regurgitant pressure gradient; %DFT = diastolic filling time.

for treatment of RV heart failure in the 10 months since the therapy (Table 1). A follow-up echocardiogram demonstrated an improvement in RV dyssynchrony (Figure 2F).

Discussion

We reported 2 patients with rTOF with RV dyssynchrony because of RBBB or RV apical pacing, in whom CRT was implemented effectively for treatment of RV heart failure refractory to medical therapy. To the best of our knowledge, this is the first report of CRT-RV by application of additional pacing at the RV free wall.

RBBB or RV apical pacing may cause electrical dyssynchrony in the RV, thus leading to RV systolic abnormalities. In previous studies, activation mapping in rTOF patients with RBBB showed delayed activation in the RV lateral wall and outflow tract relative to the septal breakthrough, similar to that in our case 1; this activation pattern causes early RV systolic abnormalities and late insufficient contraction.⁴⁻⁶ As we also observed in case 2, RV apical pacing induces an iatrogenic intraventricular delay of electrical conduction, which is reflected in interventricular and intraventricular desynchronization.⁷ Therefore, RV dyssynchrony had been identified as a therapeutic target for treatment of refractory RV heart failure.

Although an acute hemodynamic benefit of augmenting RV and systemic performance may be observed in rTOF with RBBB using biventricular or multifocal RV pacing,^{8,9} there are few studies on clinical outcomes in rTOF patients who, like our patients, specifically presented with isolated RV dysfunction and who underwent CRT. Moreover, how to improve poor RV function with CRT is a virtually unstudied area. Tambo et al reported that biventricular stimulation significantly decreased right and LV dyssynchrony and

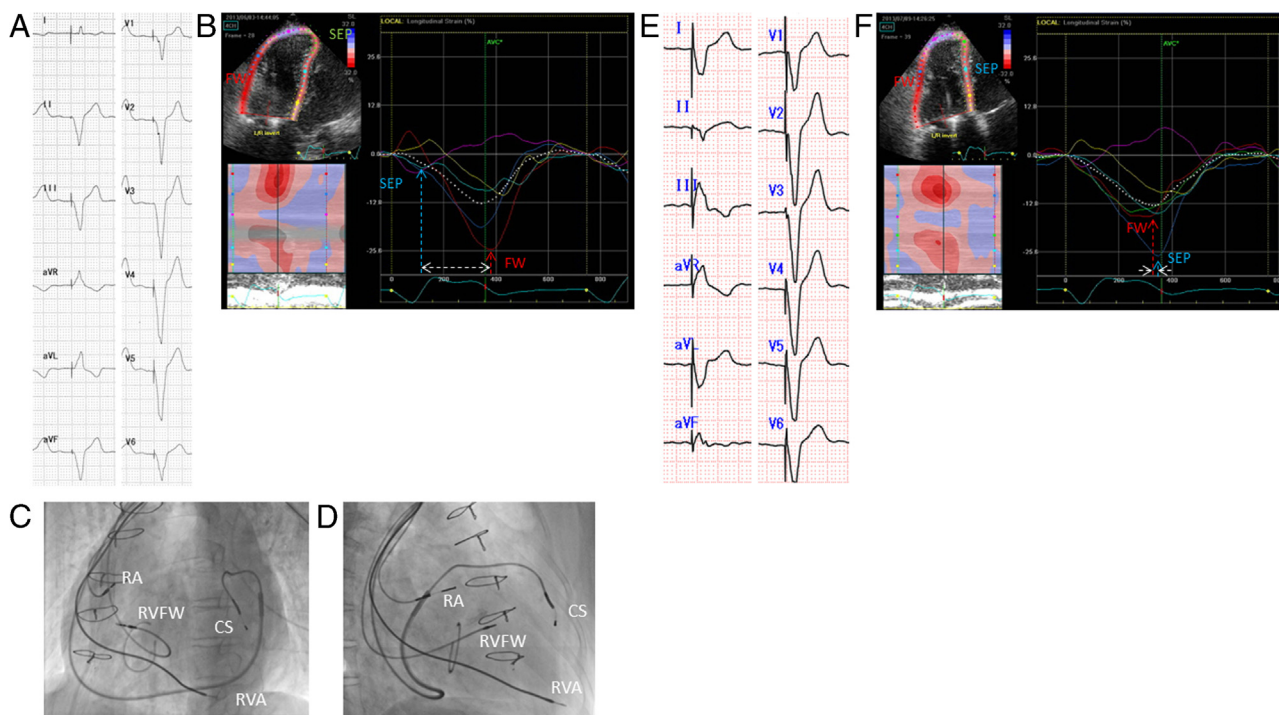


Figure 2 Data in case 2. **A and E:** 12-lead electrocardiograms before and after the implantation, respectively. QRS duration shortened from 194 to 172 milliseconds after the CRT. **B and F:** 2-dimensional speckle tracking–derived displacement curves of RV and septal segments in an apical 4-chamber view before and after the CRT, respectively. Red and blue dotted arrows indicate the negative peaks of the curves of the RVFW and mid SEP, respectively. The contraction delay of RVFW relative to the SEP was markedly improved from 259 to 0 milliseconds. **C and D:** Fluoroscopic views in left and right oblique projections showing lead positions. Other abbreviations are as in Figure 1.

improved New York Heart Association functional class at 6 months of follow-up in 9 rTOF adult patients.¹⁰ However, a delayed area in the RV, away from the RV pacing site, persisted even during biventricular pacing.¹⁰ Therefore, to create optimal resynchronization of the RV, we added another lead at the RV free wall that was the last to be activated, in addition to the RV apical lead and a left-sided lead, thus resulting in improvement in RV dyssynchrony as well as improvement of synchronized motion in the entire heart.^{11,12}

Current CRT devices are equipped with 3 ports to which the atrial, RV, and LV leads, respectively, are connected, although they are not prepared to perform pacing at ≤ 3 ventricular sites. Moreover, the atrial port is not appropriate for ventricular pacing, because the timing of pacing via the atrial port is not simultaneous to but rather 30 milliseconds or more earlier than that of pacing via other ports. Therefore, we connected the RV apical lead to the RV port to keep ventricular sensing stable, and connected the other 2 leads at the RV free wall and LV-to-LV port via the Y-connector.

One could hypothesize that standard CRT using a single RV/LV lead was the beneficial factor in both cases, obviating the need for an additional RV lead. Actually, we did not separate the additional effect of the lateral RV or LV from the net effect of CRT-RV. In addition, we did not confirm the presence of LV dyssynchrony during RV pacing. Nevertheless, we believe that the significant improvement in clinical symptoms and physiological findings due to RV failure was attributable mainly to the effect of multifocal pacing in the RV that normalized RV dyssynchrony. Although the precise mechanism of improvement in RV heart failure remains unclear, CRT-RV might promote an increase in contractility of the RV and prolongation of the diastolic phase, thus leading to improved systolic and diastolic function of the RV.

In contrast to the methodology for assessing LV dyssynchrony, the technique for assessment of RV dyssynchrony has not been established. In this study, a 2-dimensional speckle tracking technique was adapted to RV, but there is currently no evidence of a correlation with RV dyssynchrony. Nevertheless, assessments of the difference in the timing of the septal and RV free wall motion were available, and were sufficient to evaluate RV dyssynchrony and the associated improvement in clinical symptoms.

CRT-RV may be a feasible option for the treatment of RV failure in rTOF patients. Despite the better outcome reported in the present study, the necessity of implanting the LV or the additional RV lead requires further investigation. Our results are not conclusive but may provide an option to manage this type of patient. Also, further studies are needed to clarify whether this technology is effective for RV failure that is due to other etiologies.

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