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"The Heart Sound Quintet": A Case Report of Right-Sided Heart Failure Due to Free Pulmonary Regurgitation Long After Intracardiac Repair of Tetralogy of Fallot

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Data Interpretation D
Manuscript Preparation E
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Conflict of interest: None declared

Patient: Male, 52-year-old
Final Diagnosis: Tetralogy of Fallot (TOF)
Symptoms: Palpitation
Medication: —
Clinical Procedure: Computed tomography • echocardiography • magnetic resonance imaging
Specialty: Cardiology

Objective: Congenital defects/diseases

Background: Intracardiac repair of tetralogy of Fallot (TOF) is generally performed during childhood. However, the majority of patients develop the sequelae long after surgical repair, which results in significant right ventricular (RV) dilation, RV myocardial dysfunction, and, ultimately, in right-sided heart failure.

Case Report: A 52-year-old man was referred to our institution for the evaluation of sudden-onset ventricular tachycardia. His medical history included RV outflow tract reconstruction at 5 years of age. Auscultation revealed a harsh diastolic regurgitant murmur, widely split first heart sound (S_1), and a single second heart sound (S_2), indicating a severely dilated RV due to severe pulmonary regurgitation (PR) and the presence of a non-functioning pulmonary valve. Moreover, the right-sided third heart sound (S_3) and fourth heart sound (S_4) were present, consistent with elevated RV filling pressure and the presence of a non-compliant RV. Eventually, the aforementioned "heart sound quintet" was confirmed using multimodal evaluation as right-sided heart failure with a concomitant severely dilated RV because of complete regression of the pulmonary valve and resultant free PR.

Conclusions: We encountered a case with a "heart sound quintet" that was composed of a widely split S_1, single S_2 with a harsh diastolic regurgitant murmur, and right-sided S_3 and S_4. The logical interpretation of the findings from physical examination will contribute to understanding the pathophysiology and aid clinical decision-making.

MeSH Keywords: Case Reports • Heart Auscultation • Tetralogy of Fallot

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/924636

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Background

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease, accounting for 7–10% of all congenital heart diseases [1]. To date, intracardiac repair of TOF is generally performed during childhood. As a result, both structural and hemodynamic abnormalities are almost completely repaired. On the other hand, the majority of these patients will develop the sequelae long after the initial surgery, including pulmonary regurgitation (PR), right ventricular (RV) outflow tract obstruction, and pulmonary valvular stenosis, which results in significant RV dilation, RV myocardial dysfunction, and ultimately leads to right-sided heart failure [1]. Moreover, these post-operative sequelae are associated with significant morbidity and mortality for patients after intracardiac repair of TOF [2]. For these patients, high-technology imaging modalities, including cardiac computed tomography (CT), cardiac magnetic resonance imaging (MRI), and cardiac catheterization, are generally used for the assessment of structural and hemodynamic abnormalities. However, little is known regarding the utility of physical examination in the assessment of these sequelae.

Here, we present a case of repaired TOF with concomitant decompensated right-sided heart failure due to severe PR, in which the structural abnormality and hemodynamic derangements were successfully assessed in detailed physical examinations.

Case Report

A 52-year-old man was admitted to a hospital because of sudden-onset palpitation and general fatigue. Electrocardiography revealed ventricular tachycardia with a heart rate of 180 beats/min. His heart rhythm returned to a sinus rhythm after prompt cardioversion and he was then referred to our institution for further investigation. His past medical history included RV outflow tract reconstruction using a homograft and closure of a ventricular septal defect for TOF, which was performed 47 years previously. However, he was lost to follow-up thereafter. Upon referral to our institution, his blood pressure was 116/70 mmHg, heart rate 80 beats/min, and oxygen saturation of 99% (on room air). On inspection of the jugular venous pulsation, a prominent *a* wave and deep *y* descent were clearly observed, which were consistent with the increased RV filling pressure and compensated augmentation of RA contraction. Of note, auscultation revealed a harsh diastolic regurgitant murmur in the third left intercostal space, widely split first heart sound (S_1), and a single second heart sound (S_2), indicating a severely dilated RV due to free PR and the presence of a non-functioning pulmonary valve. Moreover, a third heart sound (S_3) was auscultated at the fourth parasternal border, which was consistent with a right-sided S_3. The electrocardiogram revealed complete right bundle branch block, with a QRS duration of 200 ms. Chest radiography showed cardiomegaly with dilation of the bilateral pulmonary arteries (Figure 1). Transthoracic echocardiography revealed complete regression of the pulmonary valve, which resulted in free PR and a prominent RV dilation (Figure 2, Videos 1–3). Volumetric assessment

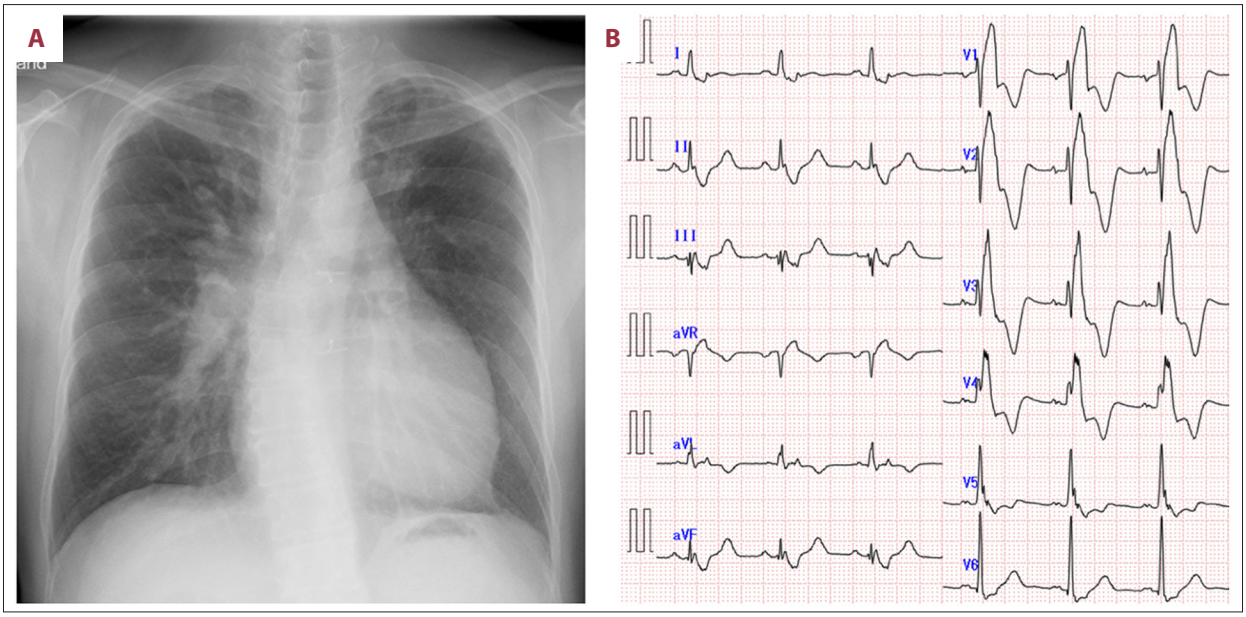

Figure 1. Chest radiograph and electrocardiogram of a 52-year-old man. Chest radiograph on admission shows significant cardiomegaly and dilation of the bilateral pulmonary arteries (**A**). Electrocardiogram on admission shows complete right bundle branch block with a QRS duration of 200 ms (**B**).

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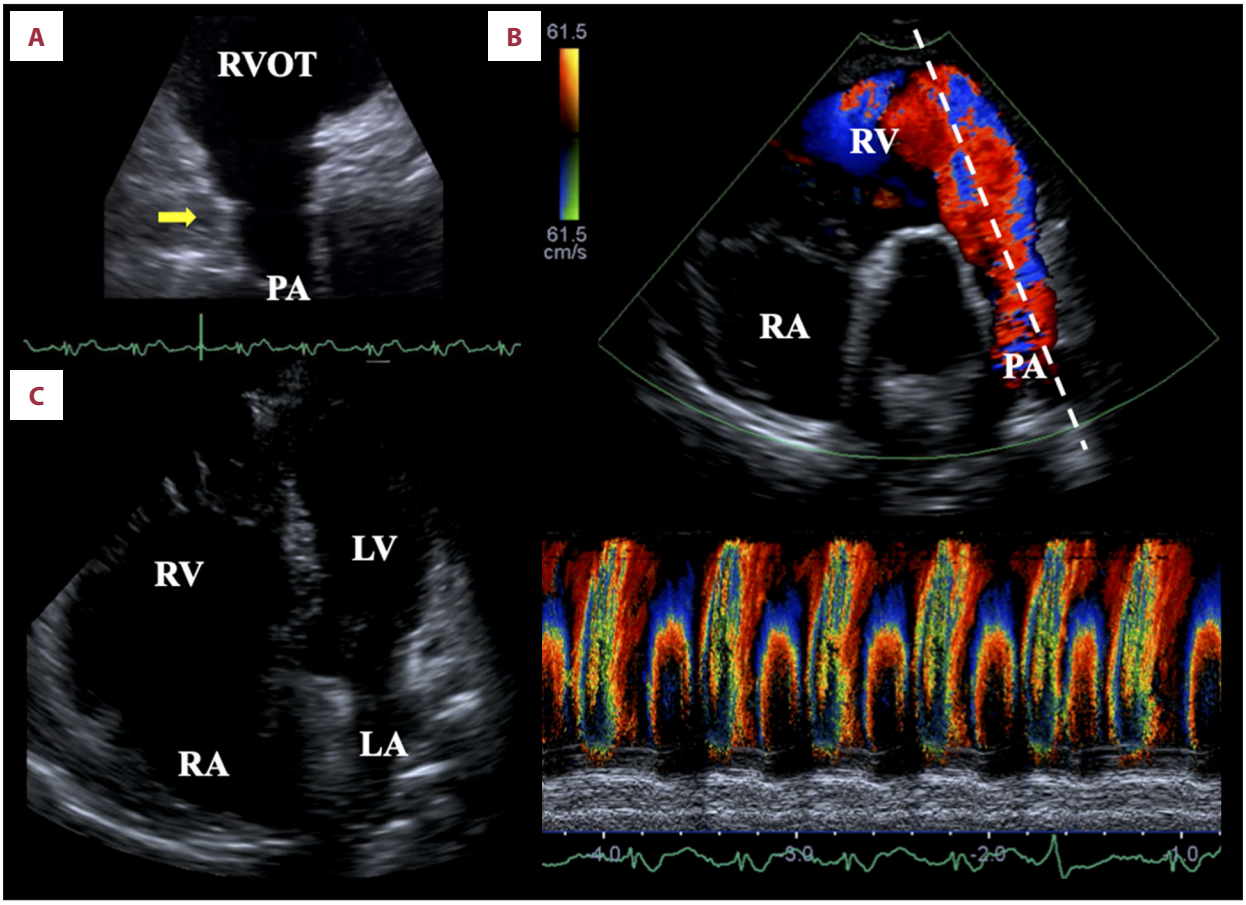
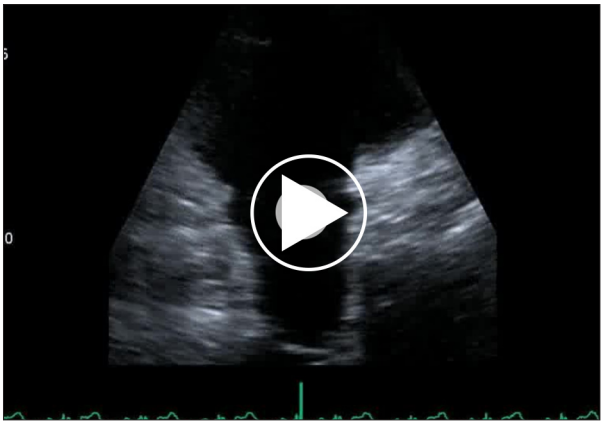
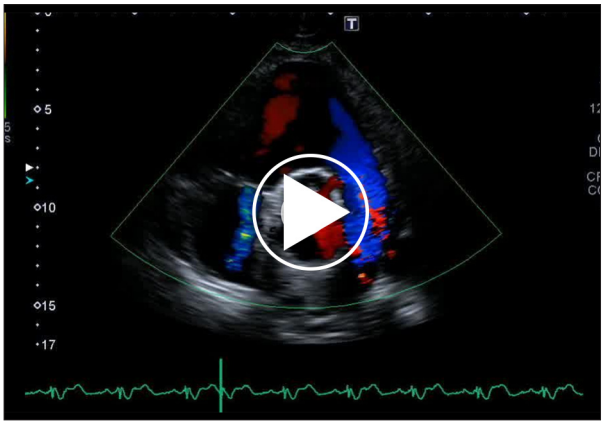


Figure 2. Transthoracic echocardiogram on admission of 52-year-old man. Parasternal right ventricular (RV) outflow view showing a complete regression of the pulmonary valve (**A**). Parasternal short-axis view shows free pulmonary regurgitation (PR). A color M-mode echocardiogram clearly shows free PR (**B**). Apical 4-chamber view shows prominent RV dilation as a result of free PR (**C**). RV – right ventricle; RVOT – right ventricular outflow tract; PA – pulmonary artery; RA – right atrium.



Video 1. Parasternal right ventricular outflow view shows the absence of the pulmonary valve.



Video 2. Parasternal short-axis view shows free pulmonary regurgitation.

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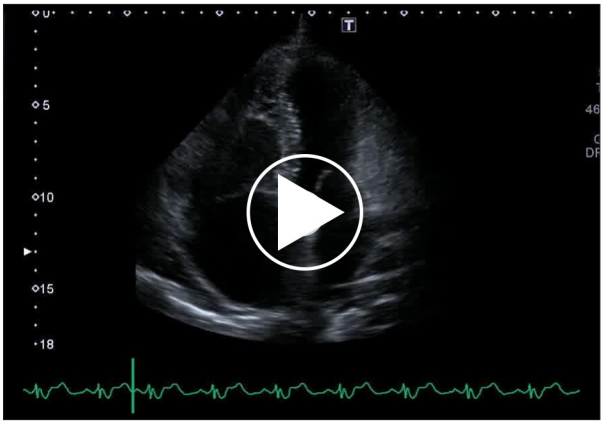
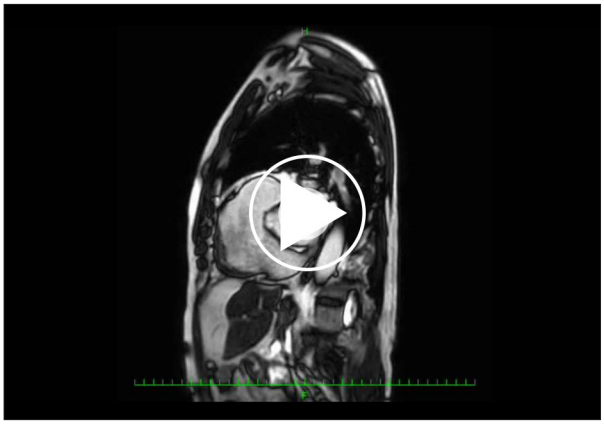


Video 3. Apical 4-chamber view shows prominent right ventricular dilation.



Video 4. Four-dimensional sagittal plane of the cardiac magnetic resonance imaging shows a remarkably dilated right ventricle and free pulmonary regurgitation.

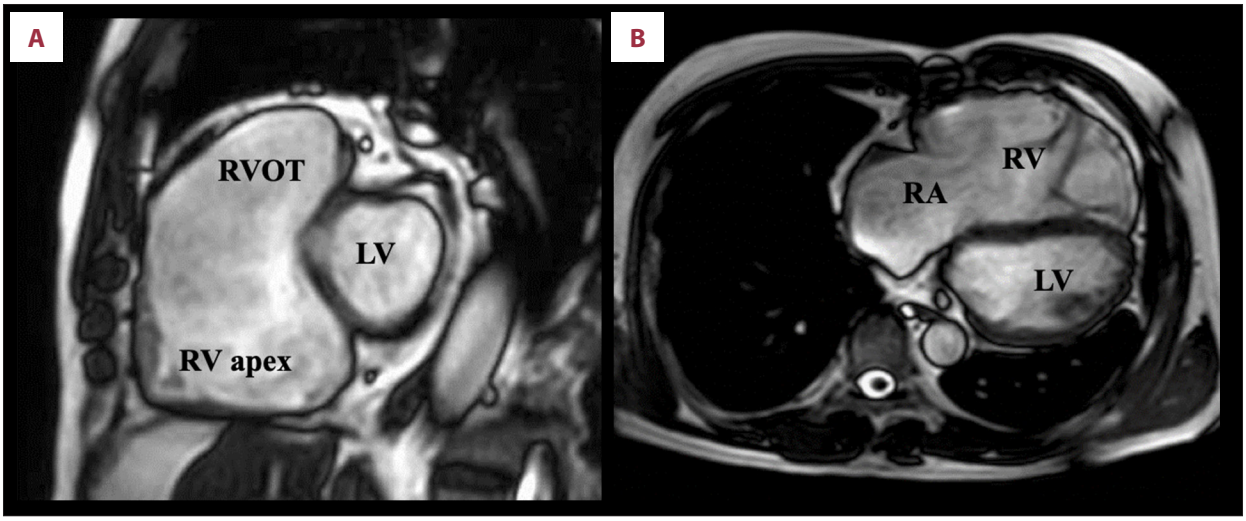


Figure 3. Cardiac magnetic resonance imaging of a 52-year-old man. Sagittal plane of the cardiac magnetic resonance imaging (MRI) shows a significantly dilated right ventricle (RV). Note the comparison with the adjacent left ventricle (**A**). Axial plane of the cardiac MRI shows the significantly dilated right atrium and RV, compressing the left side of the heart (**B**). RV – right ventricle; RVOT – right ventricular outflow tract; RA – right atrium; LV – left ventricle.

of the RV was performed using cardiac MRI, which revealed an RV end-diastolic volume of 428 ml (RV end-diastolic volume index of 240 ml/m^2), RV ejection fraction of 36%, and regurgitant fraction of PR of 74% (Figure 3, Video 4). Moreover, multi-detector row CT clearly showed complete regression of the pulmonary valve of the homograft (Figure 4).

The patient underwent catheter examination for further investigation of his hemodynamic state. His RA pressure waveform was particularly notable, consisting of a prominent *a* wave and deep *y* descent, which were coincident with the fourth heart sound (S_4) and S_3 on a phonocardiogram, respectively (Figure 5). Moreover, pulmonary arterial end-diastolic pressure and RV end-diastolic pressure were revealed to be almost identical due to the free PR. Thus the clinical assessment of the "heart sound quintet", including a harsh diastolic regurgitant murmur, widely split S_1, single S_2, and the presence of right-sided S_3 and S_4, was confirmed using multimodal anatomical and hemodynamic evaluation as severe right-sided heart failure concomitant with a significantly dilated RV because of complete regression of the pulmonary valve and resultant free PR.

After pulmonary valve replacement, the "heart sound quintet" had completely resolved.

Discussion

Hemodynamic and structural abnormalities long after intracardiac repair of TOF can result in progressive electrical remodeling

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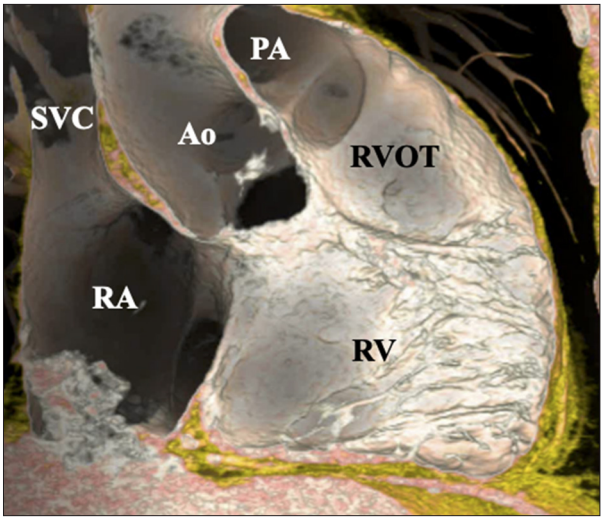

Figure 4. Multi-detector row computed tomography of a 52-year-old man. The three-dimensional volume-rendering image clearly shows complete regression of the pulmonary valve of the homograft. SVC – superior vena cava; RA – right atrium; RV – right ventricle; Ao – aorta; RVOT – right ventricular outflow tract; PA – pulmonary artery.

of the right side of the heart, and the mechano-electric interaction between QRS duration and RV dilation has been well demonstrated previously [3]. Moreover, it has been reported that RV myocardial stretch engenders areas of inhomogeneous electrical activity, predisposing to the development of life-threatening arrhythmia [3], as observed in our case.

S_1 is composed of I_M and I_T components; thus, a widely split I_M-I_T interval indicates the delay of the I_T component because of the presence of a significantly dilated RV due to hemodynamic load and could indicate a risk of life-threatening arrythmia. In this case, the RV end-diastolic volume index was revealed to be significantly dilated to 240 ml/m^2 and the I_M-I_T interval was extremely prolonged at 150 ms as measured using a phonocardiogram. Thus, a widely split S_1 can be used as a non-invasive marker of RV volume overload due to significant PR for patients after intracardiac repair of TOF.

In this case, the pulmonary valve of the homograft had almost completely disappeared due to aging degradation, resulting in free PR. Although S_2 is normally composed of II_A and II_P, in this case II_P was not auscultated because of the absence of the pulmonary valve. Therefore, a harsh holo-diastolic murmur

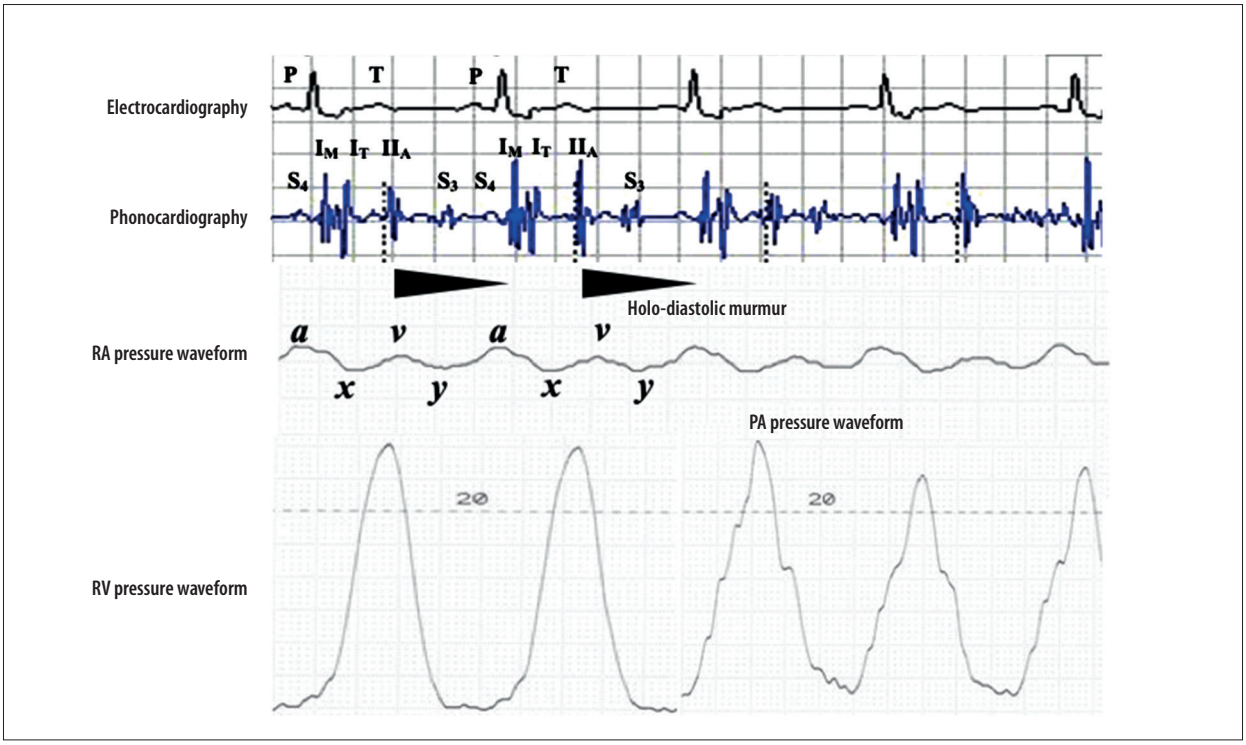

Figure 5. Simultaneous presentation of the electrocardiogram, phonocardiogram, and pressure waveforms from the right atrium, right ventricle, and pulmonary artery. The phonocardiogram clearly shows the widely split S_1, single S_2, and right-sided S_3 and S_4. Note that S_4 and S_3 are coincident with a prominent *a* wave and deep *y* descent in the right atrial pressure waveform, respectively. Pulmonary arterial and right ventricular end-diastolic pressures are almost identical due to free PR. ECG – electrocardiogram; RA – right atrial; RV – right ventricle; PA – pulmonary artery.

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with a single S_2 strongly suggested not only the presence of severe PR but also its pathogenesis.

S_3 is known to be associated with elevated ventricular filling pressure, which is generally auscultated at the apex in patients with left-sided heart failure. However, right-sided S_3 is indicated when S_3 is heard at the left parasternal border and is enhanced during inspiration, as observed in our case. In fact, in this case, the RV end-diastolic pressure was elevated to 10 mmHg and a prominent *y* descent was observed in the RA waveform, which was quite compatible with the elevated filling pressure of the right side of the heart.

Finally, S_4 is mainly created when the atrial contraction rapidly distends the ventricle in the presence of a non-compliant ventricle. Although the auscultation could not reveal the presence of S_4, S_4 was clearly exhibited on the phonocardiogram, coincident with a prominent *a* wave in the RA pressure waveform. When a massively dilated RV undergoes fibrotic remodeling and encounters space constraints due to the pericardium and thoracic cavity [4], a further increase in RV volume during atrial systole requires RA hypercontraction. Therefore, the presence of S4 is an important finding, indicating the presence of a non-compliant RV concomitant with elevated RV end-diastolic pressure.

Conclusions

We encountered a case of "heart sound quintet" that was composed of a widely split S_1, single S_2 with a harsh diastolic regurgitant murmur, and right-sided S_3 and S_4. Detailed physical examination contributed in the evaluation of not only structural but also hemodynamic derangements in a patient long after intracardiac repair of TOF. Life-long follow-up examinations in repaired TOF patients are important and will improve morbidity and mortality in this patient cohort.

Conflict of interest

None.

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