

# Treatment strategies for aorta-right ventricular fistula associated with aortic and/or mitral valve replacements: a case report with a review of the literature

Strategie leczenia przetoki między aortą a prawą komorą serca związanej z operacją wymiany zastawki aortalnej i/lub mitralnej: opis przypadku i przegląd piśmiennictwa

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## Abstract

We report a case of aorta-right ventricular fistula associated with aortic and mitral valve replacements. We considered it as a paravalvular leakage. Aorta-right ventricular fistula is a very rare variant of paravalvular leakage. We discuss the aetiology, diagnosis, and preventive and therapeutic options in the modern era.

**Key words:** aorta, right ventricle, fistula, paravalvular

## Streszczenie

Przedstawiamy przypadek pacjenta z przetoką między aortą a prawą komorą związaną z operacją wymiany zastawki aortalnej i mitralnej. Była ona rozpatrywana jako przeciek okołozastawkowy. Przetoka między aortą a prawą komorą jest bardzo rzadką odmianą przecieku okołozastawkowego. Omawiamy jej etiologię, diagnostykę oraz obecne strategie zapobiegania i leczenia.

**Słowa kluczowe:** aorta, prawa komora, przetoka, ubytek okołozastawkowy

## Introduction

Aorta-right ventricular (Ao-RV) fistulas are seen rarely. Most of these cases are caused by sinus of Valsalva rupture, endocarditis, and less frequently by trauma or are associated with valve replacement [1, 2]. We report a case of Ao-RV fistula associated with aortic and mitral valve replacements and considered as a paravalvular leakage.

## Case report

A 58-year-old male who had mechanical mitral and aortic valves was admitted to our clinic with exertional dyspnoea for 3 years. He was free of any illness rather than the previous rheumatic heart disease. His admittance heart rate was 87 per minute, arterial blood pressure was 130/70 mmHg

and the body temperature was 36.7°C. Mechanical valve sounds were audible and systolic and diastolic murmur was heard at the precordial area. The rest of the physical examinations were normal. Biochemical parameters were all in normal ranges and the haemoglobin was 11.2 mg/dl. The prothrombin time was 18 s and the international normalized ratio (INR) was 1.3, which are in the ineffective range. Electrocardiography showed normal sinus rhythm. He was operated on for severe aortic and mitral stenosis and mitral regurgitation, and the native valves were replaced by bileaflet St. Jude mechanical valves (St. Jude Medical Inc, Minneapolis, MN, USA) 3 years ago. Transthoracic echocardiography (TTE) revealed normal function of mechanical aortic and mitral valves, modest impaired systolic left ventricle function (EF 40%), dilated

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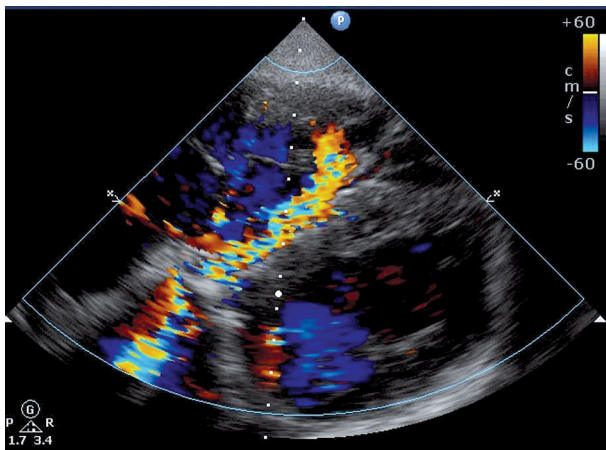
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right heart and enlarged hepatic veins. Colour Doppler revealed an abnormal jet in the right ventricle (fig. 1). Then CW Doppler revealed a systole-diastolic jet from the aorta toward the right ventricular apex (fig. 2). So we decided to perform transesophageal echocardiography (TEE) for further investigation. TEE supported the aorta-right ventricular fistula through a 20 × 8 mm, crescent, aortic paravalvular defect (fig. 3). So we performed cardiac catheterization to confirm the diagnosis and to investigate the associated pathologies. Nothing apart from the large aorta-right ventricular fistula was seen, nor was significant

coronary artery disease observed (fig. 4). We recommended him percutaneous closure of the fistula but he refused this option as well as the re-do operation, after optimizing his medical therapy he was discharged. Unfortunately he died at the ninth month of the follow-up.

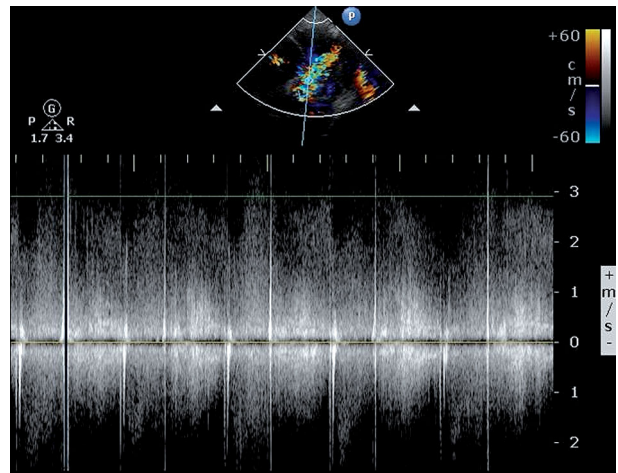
### Discussion

Aorta-RV fistulas are seen infrequently, and limited cases have been reported caused by valve replacement [2-6]. Iatrogenic injury of the septum, aggressive debridement of the annulus and inappropriate suturing of the



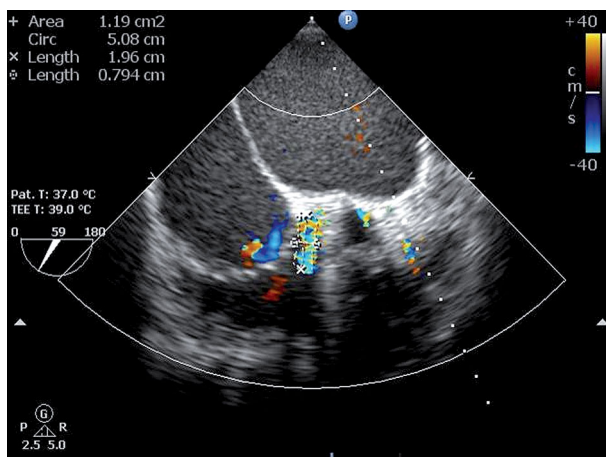
**Fig. 1.** Aorta-right ventricular fistula jet by colour Doppler from subcostal window

**Ryc. 1.** Przetoka między aortą a prawą komorą z widoczną falą przepływu w badaniu metodą dopplera kolorowego w projekcji podmostkowej



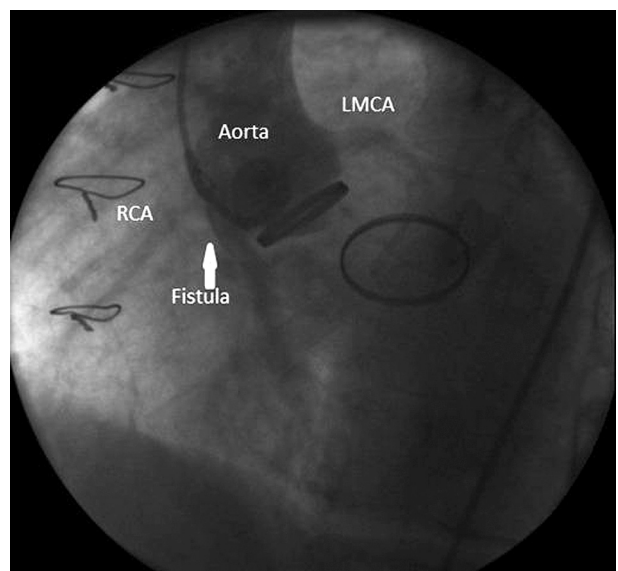
**Fig. 2.** Systole-diastolic jet by CW Doppler from subcostal window

**Ryc. 2.** Skurczowo-rozkurczowa fala przepływu w badaniu dopplerowskim fali ciągłej w projekcji podmostkowej



**Fig. 3.** Aortic paravalvular defect is seen at the right heart border of the mechanical valve annulus at 60° by transoesophageal echocardiography

**Ryc. 3.** Ubytek okotozastawkowy zastawki aortalnej widoczny na granicy prawokomorowej pierścienia zastawki mechanicznej pod kątem 60° w badaniu echokardiograficznym przezprzełykowym



**Fig. 4.** Aorta-right ventricular fistula is seen in the aortography from left anterior oblique view

**Ryc. 4.** Przetoka między aortą a prawą komorą widoczna w aortografii w projekcji przedniej skośnej lewej

mechanical valve onto the membranous portion of the ventricular septum may cause the fistula. Also primary suture failure (dehiscence) and infective endocarditis associated paravalvular leakages (PVL) may be seen [5]. Intraoperative TEE may reduce the risk of PVL, by identifying residual leaks for the surgeon to address before chest closure [7].

Presentation of aorta-RV fistulas is diverse in the aetiology, size and acute or chronic nature of the fistula. Fever may be seen in the infective states. Right heart failure may be seen in chronic phases due to RV pressure and volume overloads. Left ventricular dilatation and failure may be seen due to high output as well as in the patent ductus arteriosus or peripheral A-V fistulas [8]. Diastolic murmur may be heard in patients with native valves but metallic sounds of the mechanical valves may mask it. High gradients may be observed across the mechanical valves due to the high output states in the echocardiography. Anaemia, icterus and elevated lactate dehydrogenase level may be found because of the haemolysis [2, 9]. Diagnosis may be an incidental finding by the evaluation of left/right heart failure or routine follow-up by TTE. Once the clinician becomes suspicious of it, the diagnosis may need special attention and awareness [3]. Confirmation of the diagnosis may be done by catheterization, cardiac magnetic resonance or multi-slice computerized tomography.

The decision for the therapy is made depending on the size and clinical characteristics of the patient. Haemolytic anaemia, pulmonary hypertension, and right and/or left heart failure are the indications for treatment of the fistula. Aorta-RV fistulas and other paravalvular leakages are, conventionally, treated by open heart surgery. Experiences are limited for surgical treatment due to the infrequency of the disease. Post-operative fistulas such as paravalvular leakages have more risk due to the possible associated cardiovascular morbidities such as depressed ventricular function, pulmonary hypertension so the re-do operation [2]. Percutaneous transcatheter is a novel choice for therapy but experiences are also limited like the surgical choice. It was applied by several physicians with different devices including a coil [9], Amplatzer duct occluder [6], Amplatzer atrial septal occluder [10], Amplatzer muscular ventricular septal defect occluder [11] and Amplatzer vascular plug [12]. Most of these applications were successful although complications were also reported [13, 14].

## Conclusions

Paravalvular leakage is an important and sometimes devastating complication of mechanical valve replacement. Avoiding PVL is simpler than the treatment. Generalization

of the intra-operative TEE application to all valve replacement and other intra-cardiac repairs is the cornerstone of prevention of PVL.

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