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Transcatheter closure of perimembranous ventricular septal defect with muscular VSD occluder after infective endocarditis in a patient with previous primum atrial septal defect closure, prosthetic aortic and mitral valves replacement. (RCD code IV-2B.3)

Agnieszka Sarnecka[1]*, Piotr Weryński[2], Piotr Wilkołek[1], Lidia Tomkiewicz-Pająk[1,3], Mateusz Brózda[1], Piotr Podolec[1,3]

[1] Department of Cardiac and Vascular Diseases, John Paul II Hospital, Krakow, Poland; [2] Department of Paediatric Cardiology, Polish-American Institute of Paediatrics (PAIP), Jagiellonian University, Medical College, Krakow, Poland; [3] Centre for Rare Cardiovascular Diseases, John Paul II Hospital, Krakow, Poland

Abstract

Ventricular septal defect (VSD) is the most common congenital heart defect at birth but is relatively rare in adult population. Apart from congenital, VSD may also be acquired. It can be caused by trauma, myocardial infarction or previous cardiac surgeries such as valvular replacements or VSD closure attempts. Surgical closure of VSD remains the treatment of choice while transcatheter closure could be considered in selected patients. Herein, we present a case of a 53-year-old man with symptomatic perimembranous VSD after infective endocarditis and a history of previous primum atrial septal defect closure and prosthetic aortic and mitral valves replacement. The VSD was successfully percutaneously closed across the prosthetic aortic valve. JRCD 2017; 3 (2): 59–64

Key words: rare cardiovascular disease, acquired ventricular septal defect, transcatheter intervention

Background

Ventricular septal defect (VSD) is the most common congenital heart defect at birth but is relatively rare in adult population. Apart from congenital, VSD may also be acquired. It can be caused by trauma, myocardial infarction or previous cardiac surgeries such as valvular replacements or VSD closure attempts. Surgical closure of VSD remains the treatment of choice while transcatheter closure could be considered in selected patients [1–4].

The shunt through a VSD leads to left ventricular (LV) volume overloading and eventually to heart failure (HF). Interventional treatment of VSD is indicated in patients with symptoms or evidence of LV volume overload, who have no severe pulmonary vascular disease [European Society of Cardiology (ESC) class I recommendation]. It should be also considered in patients with a history of infective endocarditis, VSD-associated prolapse of aortic valve cusp causing progressive aortic regurgitation as well as in patients with VSD and pulmonary arterial hypertension, if left-to-right shunt ratio (Qp:Qs) is still >1.5 and pulmonary arterial pressure (PAP) or pulmonary vascular resistance (PVR) are <2/3 of systemic values (ESC class IIa recommendation). However, according to the guidelines surgical closure of VSD remains the treatment of choice while transcatheter closure may be considered in patients at increased risk for surgery, after multiple previous cardiac surgeries or poorly accessible VSD for surgical closure as an alternative therapeutic option [1,2].

Conflict of interest: none declared. Submitted: January 21, 2017. Accepted: February 27, 2017.

* Corresponding author: Department of Cardiac and Vascular Diseases, John Paul II Hospital, Krakow, Poland, Pradnicka str. 80, 31-202 Krakow, Poland; tel. 0048 12 614 22 87, fax 0048 12 423 43 76; e-mail: a.sarnecka@szpitaljp2.krakow.pl

Case presentation

A 53-year-old Caucasian man with a history of previous cardiac surgeries was referred to the Department of Cardiac and Vascular Disease in March 2016 due to VSD after infective endocarditis for detailed cardiologic evaluation.

At the age of 20 he was diagnosed with primum atrial septal defect, which was closed surgically. The second surgery was performed at the age of 50 because of infective endocarditis complications and included mechanical mitral (St. Jude Medical 27 M) and aortic (St. Jude Medical 21 A) valves replacements.

Other concomitant conditions involved persistent atrial flutter (AFl), chronic HF with preserved ejection fraction and multiple cardiovascular risk factors such as obesity, arterial hypertension, hypercholesterolemia and diet-treated type 2 diabetes mellitus. He had also a history of central left retinal artery occlusion.

The diagnosis of acquired VSD was first established in January 2016 when the patient was admitted to a general hospital because of AFL of unknown duration with heart rate of 130–160 beats per minute (bpm). At that time the patient complained of shortness of breath in class III by New York Heart Association (NYHA) but denied fever or other symptoms of infection. Blood tests revealed elevated inflammatory markers and positive blood cultures (Staphylococcus epidermidis). Transthoracic echocardiography (TTE) did not show any pathology of prosthetic or native valves but revealed VSD with left-to-right shunt. Infective endocarditis was treated with vancomycin and ciprofloxacin for 4 weeks according to the antibiogram. Moreover, pharmacotherapy of chronic HF and rate control strategy of AFL was implemented.

On admission to our Centre the patient still complained of easy fatigue and low exercise tolerance – NYHA class III. Physical examination revealed no signs of HF. Electrocardiogram showed AFl with heart rate of 100 bpm.

Laboratory workup showed slightly elevated creatinine and high-sensitivity C-reactive protein (hsCRP), normal level of white blood cells (WBC) and therapeutic international normalized ratio (INR) level for warfarin treatment (Table 1).

TTE showed enlargement of both atria and the right ventricle (RV), paradoxical intraventricular septal motion, normal size and ejection fraction of the LV (LVEF 55%), normal prosthetic mitral and aortic valves function, moderate tricuspid regurgitation and perimembranous VSD with left-to-right shunt and approximate pressure gradient across the VSD of 90 mm Hg (Table 2, Figure1). Transesophageal echocardiography (TEE) also showed no signs of endocarditis or prosthesis valves dysfunction. More precise evaluation of VSD morphology and its relation to adjacent structures was performed using cardiac computed tomography. The study showed VSD sized 11 × 6 mm and located at a distance of 5 mm to mitral valve prosthesis and 9 mm to aortic valve prosthesis (Figure 2).

Right heart catheterization confirmed left-to-right shunt with a Qp/Qs ratio of 1.91. Cardiac output was 3.59 l/min and cardiac index 1.78 l/min/m^2. PAP was elevated and PVR was within normal limits (Table 3 and 4). Coronary angiography revealed no stenotic lesions in coronary arteries.

The patient was qualified for percutaneous closure of VSD because of high risk of surgical treatment and favorable morphology

Table 1. Laboratory workup

blood test	value	unit	reference values
WBC	6.99	10^3/mcL	[3.80–10.00]
RBC	4.77	10^6/mcL	[3.70–5.10]
HGB	14.1	g/dL	[12.0–16.0]
HCT	44.2	%	[37.0–47.0]
MCV	92.7	fL	[80.0–99.0]
PLT	200	10^3/mcL	[140–440]
INR	3.8		[0.8–1.2]
hsCRP	11.5	mg/L	[<3.0]
Creatinine	136	µmol/L	[62–106]
eGFR	51	mL/min/1,73 m^2	[>60]
K+	4.7	mmol/L	[3.5–5.1]
Na+	142	mmol/L	[136–145]
TSH	1.20	µmol/L	[0.27–4.20]
Glucose	6.6	mmol/L	[3.4–5.6]
ALT	24	U/l	[<33]
AST	32	U/l	[<23]

WBC – white blood cells, RBC – red blood cells, HGB – hemoglobin, HCT – hematocrit, MCV – mean cell volume, PLT – platelets count, INR – international normalized ratio, hsCRP – high-sensitivity C-reactive protein, eGFR – estimated glomerular filtration rate, TSH – thyroid stimulating hormone, ALT – alanine aminotransferase, AST – aspartate transaminase

Table 2. Selected transthoracic echocardiography parameters before and 2 months after VSD percutaneous closure

parameter	unit	Before VSD closure	2 months after VSD closure
LV	mm	48/32	49/32
LA	cm^2	45	33
RVOT prox	mm	41	34
RVD1	mm	53	46
RA	cm^2	38	22
TR		moderate	mild
RVSP	mm Hg	43+15	21+8
TAPSE	mm	18	16

LV – left ventricle, LA – left atrium, RV – right ventricle, RVOT prox – proximal RV outflow tract, RVD1 – basal RV dimension, RA – right atrium, IVS – intraventricular septum, TR – tricuspid regurgitation, RVSP – RV systolic pressure, TAPSE – tricuspid annular plane systolic excursion

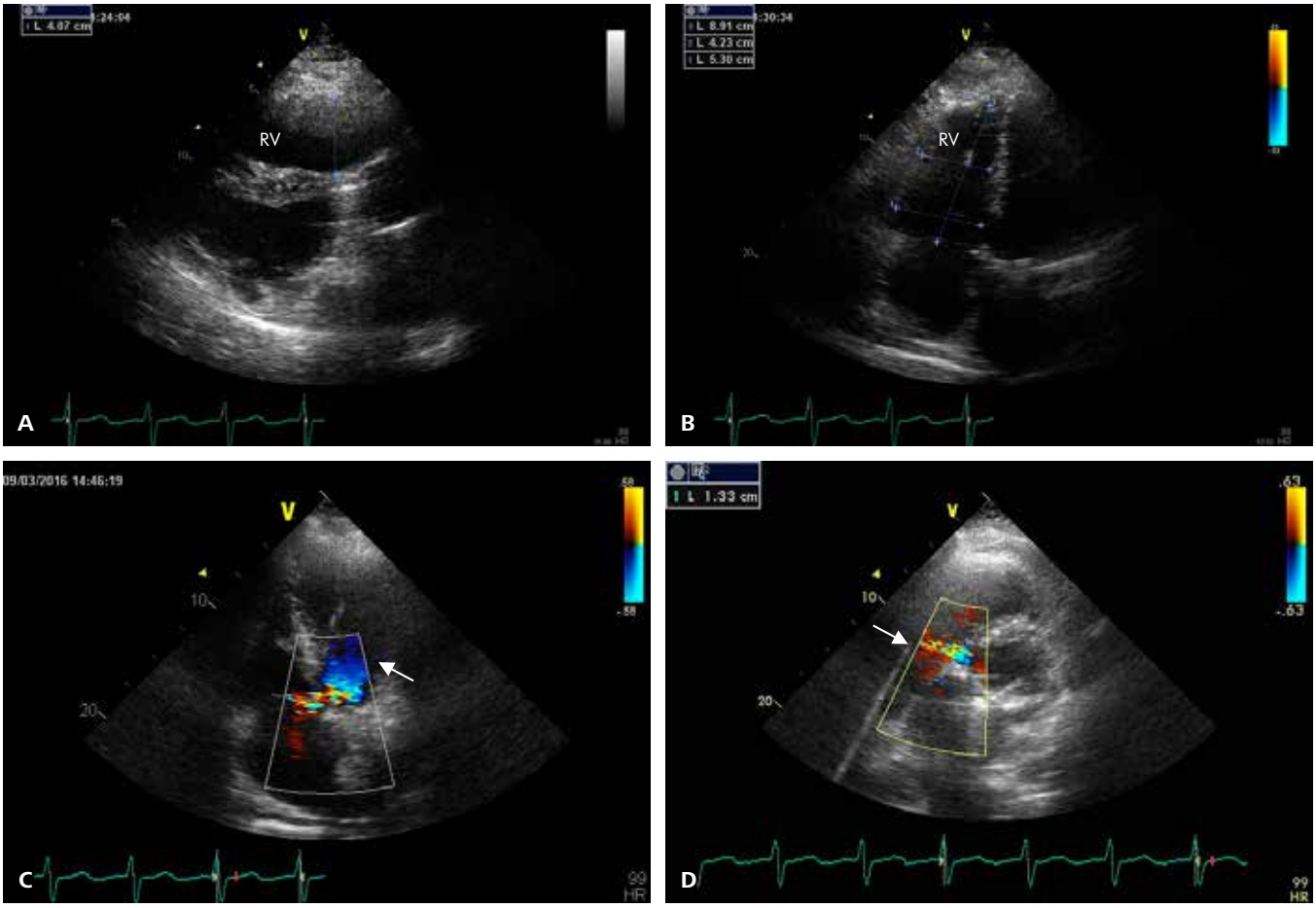

Figure 1. Transthoracic echocardiography before VSD closure showed enlarged size of both atria and right ventricle (RV) and perimembranous ventricular septal defect with left-to-right shunt (arrows). **A.** Parasternal long axis view. **B, C.** Apical four-chamber view. **D.** Parasternal short axis view

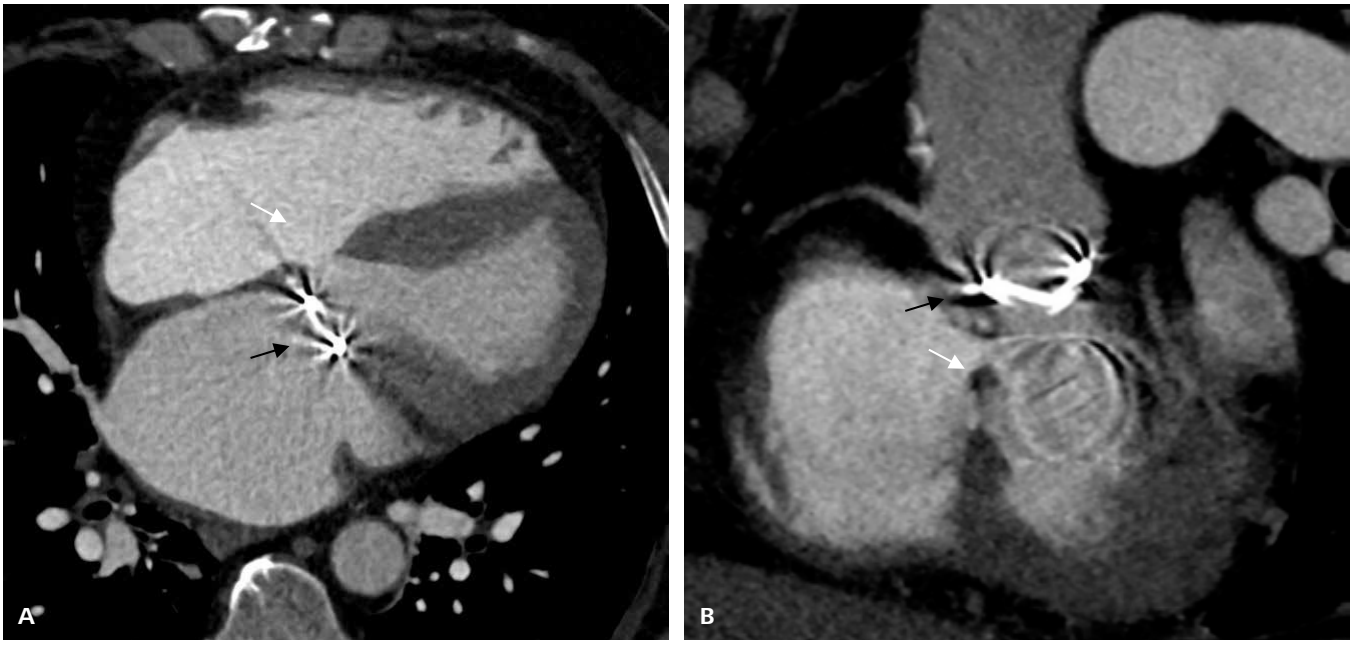

Figure 2. Computed tomography angiography of the heart showed VSD (white arrows) size of 11 x 6 mm and location at a distance of 5 mm to mitral valve prosthesis (**A.** black arrow) and 9 mm to aortic valve prosthesis (**B.** black arrow)

Table 3. The results of right heart catheterisation before VSD percutaneous closure – pressures and oxygen saturation

measurement point	pressure [mm Hg]	oxygen saturation [%]
aorta	125/94, mean 99	96
PA	63/40, mean 48	77
RV	61/4, mean 5	79
RA	/28, mean 21	77
PCW	31/40, mean 33	96
SVC		56
IVC		67

PA – pulmonary artery, RV – right ventricle, RA – right atrium, PCW – pulmonary capillary wedge, SVC – superior vena cava, IVC – inferior vena cava

Table 4. The results of right heart catheterisation before VSD percutaneous closure – resistances

measurement point	resistance [ARU]
PVR	173
TPR	554
SVR	1738
TSR	2206

PVR – pulmonary vascular resistance, TPR – total pulmonary resistance, SVR – systemic vascular resistance, TSR – total systemic resistance

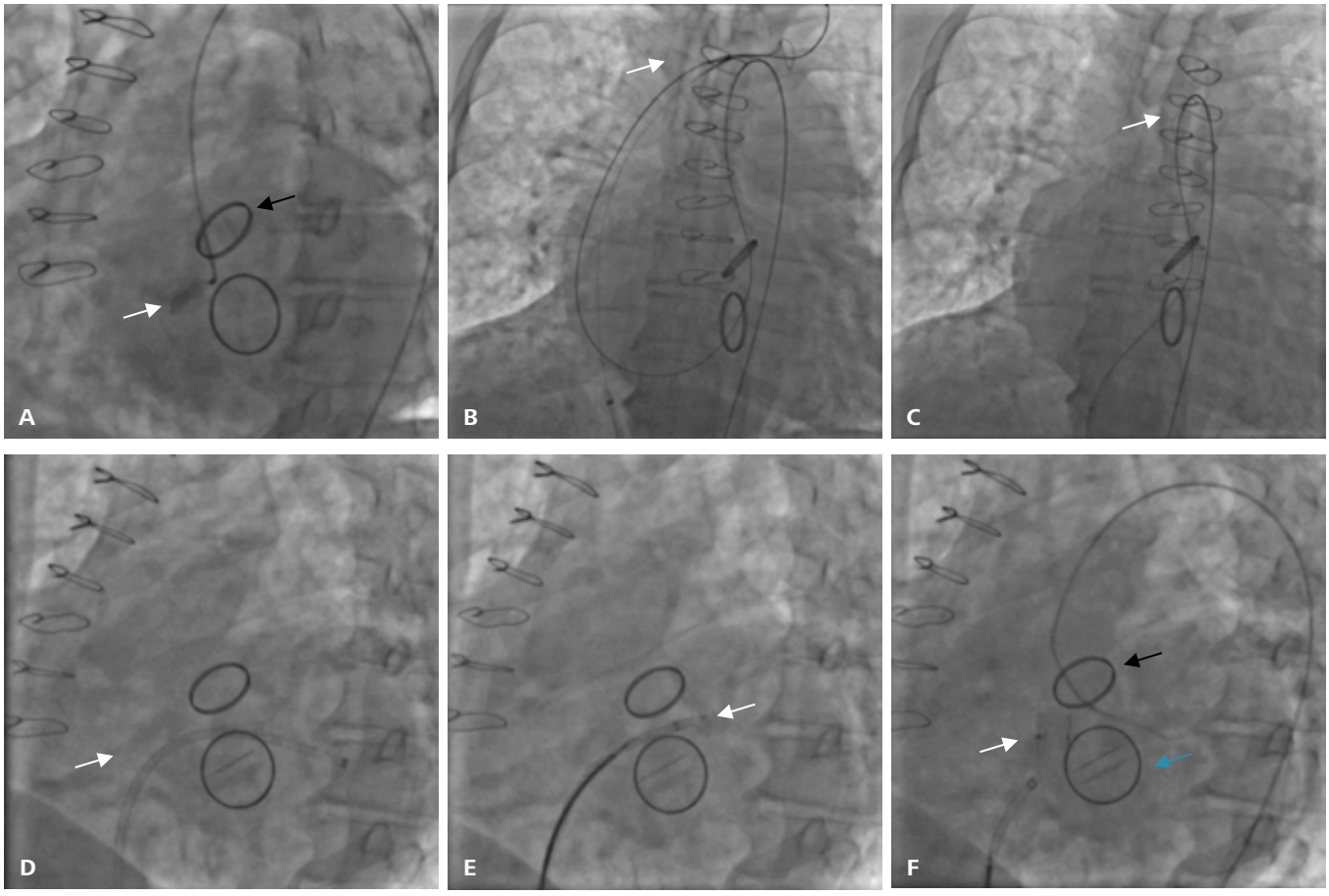

Figure 3. Cine angiographic images demonstrating steps of transcatheter closure of VSD. **A.** Retrograde left ventricle (LV) angiogram through the prosthetic aortic valve (black arrow) demonstrating the perimembraous VSD (white arrow). **B.** Snaring the guidewire advanced to right ventricle (RV) across the VSD to form an arteriovenous loop (arrow). **C.** Formatting an arteriovenous loop (arrow) after externalization of a guidewire. **D.** Transferring a delivery sheath (arrow) through the VSD from the RV to LV. **E.** Releasing the distal disc of the device from the LV (arrow). **F.** Opening the proximal disc form the RV and releasing the device (white arrow). Note the relationship between the device and prosthetic aortic (black arrow) and mitral (blue arrow) valves

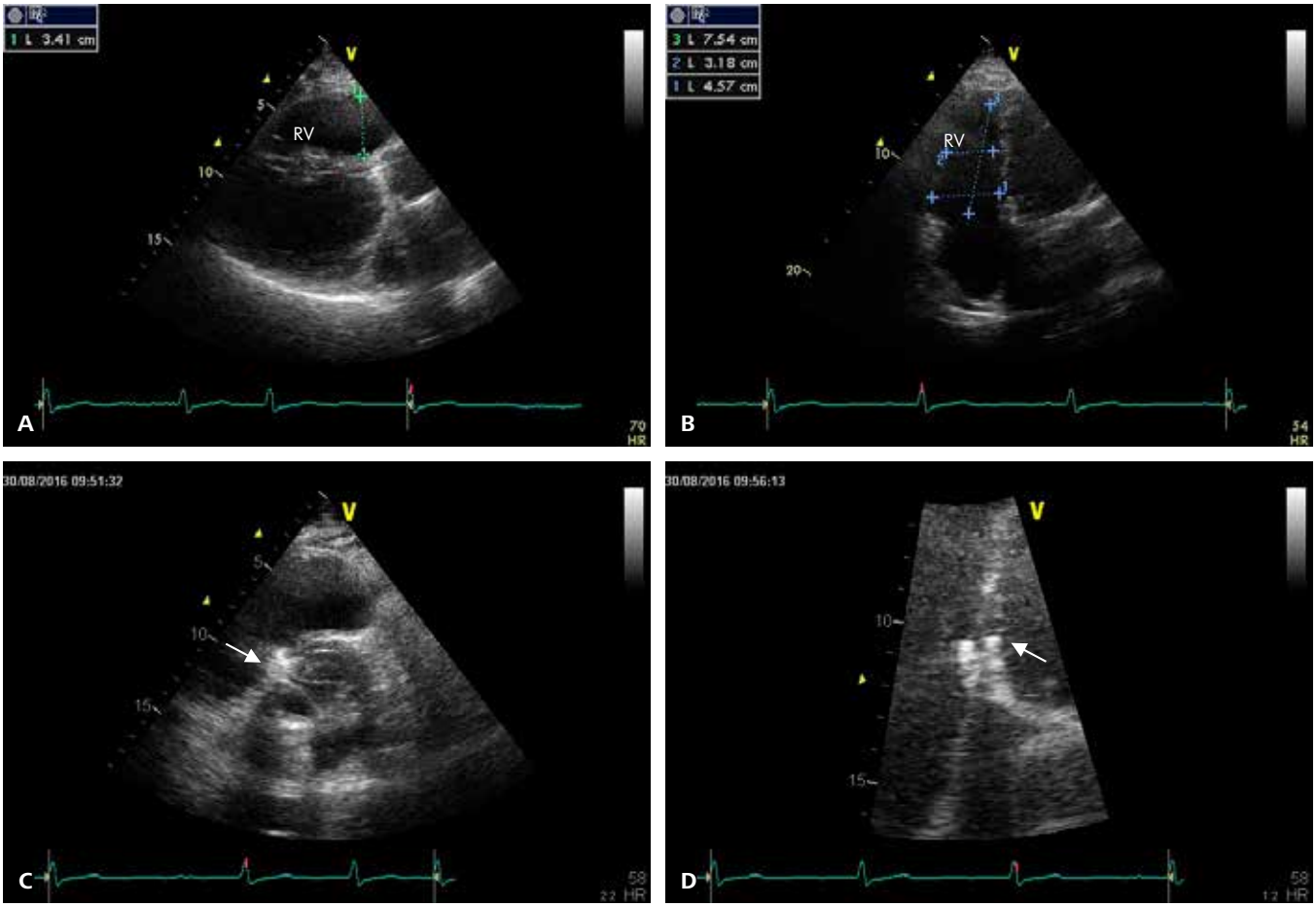

Figure 4. Transthoracic echocardiography performed 2 months after closure showed correct position of the VSD occluder (arrow) without shunt and decrease in size of right ventricle (RV) and right atrium. **A.** Parasternal long axis view. **B, D.** Apical four-chamber view. **C.** Parasternal short axis view

of the VSD. However, the risk of percutaneous procedure remained high.

VSD closure was performed with the guidance of TTE (VSD was invisible in TEE due to artifacts caused by the prosthetic valves) and fluoroscopy, in the long axial oblique view, with the patient under local anaesthesia. The first attempt to cross the VSD with a guidewire from the RV was unsuccessful. Therefore, VSD was crossed from the LV through the prosthetic aortic valve with a BER 5F catheter and Multi Snare® VSD Loop Set guidewire, that were advanced into the RV and then to the pulmonary artery. The catheter was retracted because of temporary blockage of aortic prosthetic valve discs accompanied by circulatory instability. The "kissing catheter" technique was not possible to apply. The guidewire was snared in pulmonary artery and externalized via the right femoral vein to create an arteriovenous loop. Using only guidewire a 6F AMPLATZER™ TorqVue™ delivery sheath was advanced via the right femoral vein but the first attempt to place the AMPLATZER™ VSD Occluder 10 mm across the VSD failed due to its instability and prolapse to the RV. The second attempt taken with a BER 6F catheter snaring in vena cava superior was successful. First deployment of LV disc was performed and then the entire system was pulled back against the membranous septum. The muscular AMPLATZER™ VSD Occluder was used to close perimembranous VSD because this type of device has smaller LV disc then the perimembranous occluder. This choice seems to be the best when the distance from VSD to aortic valve prosthesis is small as it was in our patient. In positioning the device from the LV, the mobility of prosthetic aortic valve discs was checked and both collision, and touching between occluder and prosthesis were excluded. Both TTE and ventriculography confirmed correct position of the VSD occluder, small residual shunt through the device and normal aortic prosthesis discs motion. (Figure 3).

Postprocedural period was complicated by recurrent pseudoaneurysm of right femoral artery, that was first treated with ultrasound-guided thrombin injection but eventually required surgical repair. Additionally, two days after the procedure patient presented symptoms of acute cholelithiasis with an increase of CRP up to 288 mg/l. Initial conservative treatment and empiric antibiotic therapy with ciprofloxacin was introduced and the patient underwent endoscopic cholecystectomy two months later. Follow-up TTE and TEE studies excluded endocarditis and showed excellent result of the VSD closure.

Review of literature

Hemodynamically significant VSD following aortic valve replacement is a rare but important complication. Surgery is still considered as the gold-standard treatment in patients with VSD but is associated with a higher surgical risk in patients after prior sternotomy. Therefore, percutaneous approach is an attractive alternative to surgical treatment in selected cases. [1,2]

The first successful percutaneous VSD closure was performed in 1988 and since then both the technique and devices have improved significantly. Preferred type of VSD for transcatheter procedure are muscular ones, that account for 15–20% cases. Device closure of much more common perimembranous defects, accounting for about 80% of VSDs, is also feasible but is associated with increased risk of atrioventricular blocks or valve damage. However, in the published case series regarding perimembranous VSDs closure with muscular VSD occluders, atrioventricular blocks were not observed. The risk of these major complications and the success rate were reported to be similar in comparison to surgical procedures [1,2,5–9].

Transcatheter closure of perimembranous VSD usually requires retrograde approach. The access to the LV across a prosthetic aortic valve during cardiac catheterization is associated with a risk of catheter entrapment, disruption of the prosthetic valve and even death. To avoid blocking the mechanical aortic valve discs when the "kissing catheter" technique is not possible to use, VSD occluder deployment can be achieved using only a guidewire. Reports of successful percutaneous closure of iatrogenic VSDs after aortic valve replacement, that have been published in the past few years are encouraging [10–15].

We conclude, that the most likely cause of VSD in our patient was infective endocarditis. The patient was at high risk of surgical treatment because of previous surgical primum atrial septal defect closure and prosthetic aortic and mitral valves replacement. However, location of the defect next to previously implanted structures and the necessity of retrograde approach made the percutaneous intervention challenging.

Patient management and follow-up

Two months after the procedure our patient was asymptomatic. TTE showed correct position of the VSD occluder without residual shunting, normal prosthetic valves function as well as decreased in size of the right heart chambers (Table 3 and Figure 4). ECG Holter monitoring demonstrated AFL with mean heart rate of 86 bpm (minimum of 59 bpm, maximum of 122 bpm).

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