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CASE REPORT
Other

More in, better out? Successful valve-in-valve procedure of an iatrogenic ventricular septal defect following transcatheter aortic valve replacement: a case report

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Background A rare, but serious, complication following transcatheter aortic valve replacement (TAVR) is the occurrence of an iatrogenic ventricular septal defect (VSD).

Case summary We describe a case of an 80-year-old female who was referred with severe aortic stenosis for TAVR. Following thorough evaluation, the heart team consensus was to proceed with implantation via a transapical approach of an ACURATE neo M 25 mm valve (Boston Scientific, Natick, MA, USA). The valve was deployed harnessing transoesophageal echocardiographic (TOE) guidance under rapid pacing with post-dilation. Directly afterwards a very high VSD close to the aortic annulus was detected. As the patient was haemodynamically stable, the procedure was ended. The next day another TOE revealed a shunt volume (left-to-right ventricle) between 50% and 60%. Because the defect was partly located between the stent struts of the ACURATE valve decision was made to fix this leakage with implantation of a further valve and we chose an EVOLUT Pro 29 mm (Medtronic Inc., Minneapolis, MN, USA). The valve-in-valve was implanted 2–3 mm below the lower edge of the first valve, more towards the left ventricular outflow tract (LVOT) with excellent result: VSD was reduced to a very small residual shunt without any hemodynamic relevance.

Discussion We suggest that an iatrogenic VSD located near the annulus may be treated percutaneously in a bail-out situation with implantation of a second valve that should be implanted slightly more into the LVOT to cover the VSD.

Keywords TAVR • Aortic annulus rupture • Iatrogenic ventricular septal defect • Valve-in valve • Case report

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Learning points

- Aortic annulus rupture is a serious complication in transcatheter aortic valve replacement (TAVR). In rare cases, the annulus ruptures into the right ventricle, which may result in a ventricular septal defect (VSD).
- Iatrogenic VSDs should be considered a rare complication of TAVR and must be differentiated from paravalvular leakage.
- A valve-in-valve (VIV) procedure that implants a second valve into the left ventricular outflow tract may be a solution for such patients in a bail-out situation.
- A self-expanding valve is preferable in cases of VIV implantation to fix a high VSD and avoid further enlargement of the defect. A beneficial effect might result from the outer skirt and the smooth-edged shape of the VIV.

Introduction

Transcatheter aortic valve replacement (TAVR) is a well-established treatment option for symptomatic, severe aortic stenosis in the elderly or patients at medium and high risk of mortality from surgical aortic valve replacement.[1] While the number of serious complications involved with TAVR has decreased, there remain life-threatening complications such as rupturing of the aortic annulus, perforation with tamponade, stroke, myocardial infarction, and serious bleeding. Other problems include the occurrence of high-grade atrioventricular (AV) blocks, paravalvular leakage (PVL), and the uncertain durability of the valves. A rare, but serious, complication is an iatrogenic, sub-annular, ventricular septal defect (VSD). Rupture of the aortic annulus or a VSD often leads to conversion to open surgery. In the case presented here, we describe a less invasive bail-out option for iatrogenic VSD treatment.

Case presentation

An 80-year-old woman in good general condition (160 cm, 60 kg) with severe aortic stenosis was admitted to our centre for TAVR. She had a history of renal insufficiency, hypertension, hypercholesterolaemia, persistent atrial fibrillation requiring oral anti-coagulants, and chronic obstructive pulmonary disease. She complained of progressive dyspnoea [New York Heart Association (NYHA) III] with evidence of cardiac decompensation (proBNP = 1410 pg/mL). Aortic stenosis was confirmed by echocardiography (mean transvalvular gradient = 45 mmHg; peak aortic jet velocity on continuous wave Doppler ultrasound = 4.3 m/s in normal left ventricular function; left ventricular ejection fraction = 65%). Additional echocardiograph findings included mild mitral and tricuspid regurgitations and moderate pulmonary hypertension (systolic pulmonary arterial pressure = 61 mmHg plus central venous pressure). A physical examination showed a typical systolic heart murmur, but no oedema or other signs of cardiac decompensation. Pre-procedure electrocardiogram documented sinus rhythm with a heart rate of 56 b.p.m. and first-degree AV block. Previous medication included amiodarone, rivaroxaban, the common antihypertensive medication, and low-dose steroids. Coronary artery disease was excluded by invasive angiogram.

Timeline

Day	Event
24 February 2020	Admission for transcatheter aortic valve replacement (TAVR)
26 February 2020 (procedure Day 0)	Transapical TAVR procedure
Day 2	Transfemoral TAVR procedure for the treatment of a ventricular septal defect (VSD) (valve-in-valve); implantation of a pacemaker
Day 3	Extubation
Day 5	Transfer to an intermediate care ward
Day 7	Pleural drainage removed
Day 16	Urinary tract infection, antibiotic therapy administered
Day 20	Beginning of thoracic wound inflammation, antibiotic and local therapy administered
Day 22	Pleuracentesis and insertion of a Mattys pleural catheter, which was removed following day
Day 26	Inflammation of the thoracic wound without improvement: change of antibiotic therapy
Day 42	Transfer to a regular ward
Day 48	Hospital discharge
May 2020	Follow-up echocardiogram: normal left ventricular function, small VSD, mean gradient of 8 mmHg
July 2020	Follow-up telephone contact: patient recovered well and had returned to normal life activities, including driving and shopping

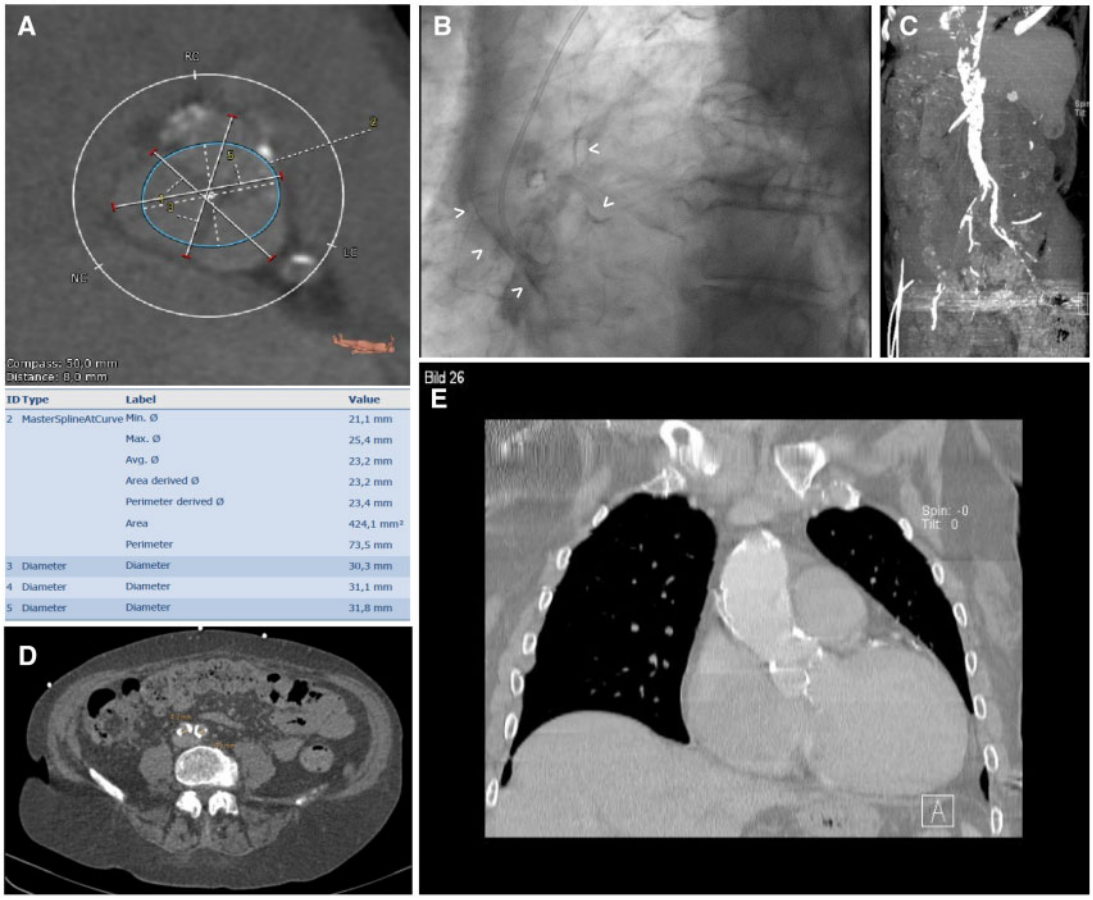

Figure 1 (*A*) 3Mensio evaluation of a pre-intervention computed tomography scan; (*B*) native radiography in the right anterior oblique projection showing the porcelain ascending aorta (arrowheads). Computed tomography scan illustrating severe calcification of the entire aorta (*C*) and the calcification of the iliac arteries with narrowing (*D*). (*E*) Computed tomography scan demonstrating severe calcification of the ascending aorta, bulbus, and the aortic arch.

Further imaging evaluation via computed tomography (CT) revealed an annulus diameter of 23.2 mm and a bulbus diameter of 31 mm (*Figure 1A*). The scan also showed a massive calcified aorta that included the subclavian and coronary arteries. The aortic annulus was moderately calcified, while the valve's leaflets were significantly calcified (*Figure 1B*, *C*, and *E*). A consensus was arrived at to perform TAVR after taking into consideration the aortic calcification, a score of 9.3% on the EuroSCORE II and the patient's age. Due to the calcifications of nearly all the vessels and moderate calcification of the valve, a self-expanding valve was deemed appropriate. Furthermore, given the supra-annular design and large valve opening area of these valve types, there would be a lower risk of annulus rupture. As there was severe calcification in both iliac arteries, with lumen reduction (minimal diameter = 4.5 mm), transapical access was preferred (*Figure 1C* and *D*). Moreover, the surgeons from our cardiac team are experienced in transapical access and have had good results.[2,3] Written consent was obtained from the patient before the procedure.

A self-expanding ACURATE neo M 25 mm valve (Boston Scientific, Natick, MA, USA) was implanted transapically with predilation (Osypka 25 × 40 mm; filled 32 mL, used 28 mL) under general anaesthesia and with transoesophageal echocardiographic (TOE) guidance. After release of the prosthesis, there was mild PVL, which was observable at the grade I–II levels. Therefore, a post-dilation was performed as the radial force of the ACURATE neo valve is known to be weak and a PVL greater than grade I is associated with higher mortality.[4] Following post-dilation (Osypka 25 × 40 mm; filled 32 mL, used 30 mL), a moderate PVL (*Figure 2A*) and a significant VSD with a jet from the annulus into the right ventricle was visible in an intraprocedural TOE (*Figures 2B* and *3B*).

Given the complete calcification of the ascending aorta and aortic arch (*Figure 1B*, *C*, and *E*), conversion to conventional surgery was rejected. As the patient was haemodynamically stable, we decided to end the procedure. The next day, another TOE was performed that revealed a shunt volume (left-to-right ventricle) between 50% and 60%. This was a membranous VSD and partly between the stent

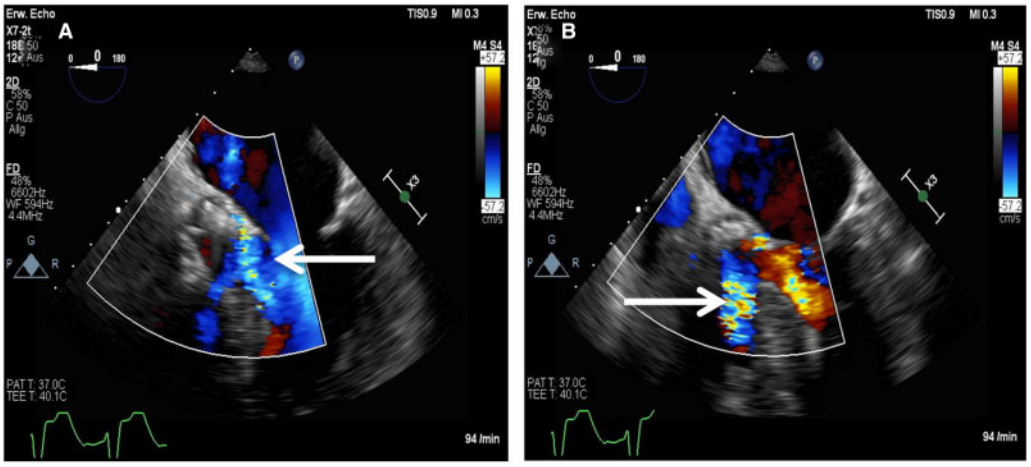

Figure 2 Transoesophageal echocardiography after implantation of a self-expanding ACURATE neo M 25 mm valve with (*A*) moderate paravalvular leakage (two small jets) and (*B*) a new, high ventricular septal defect.

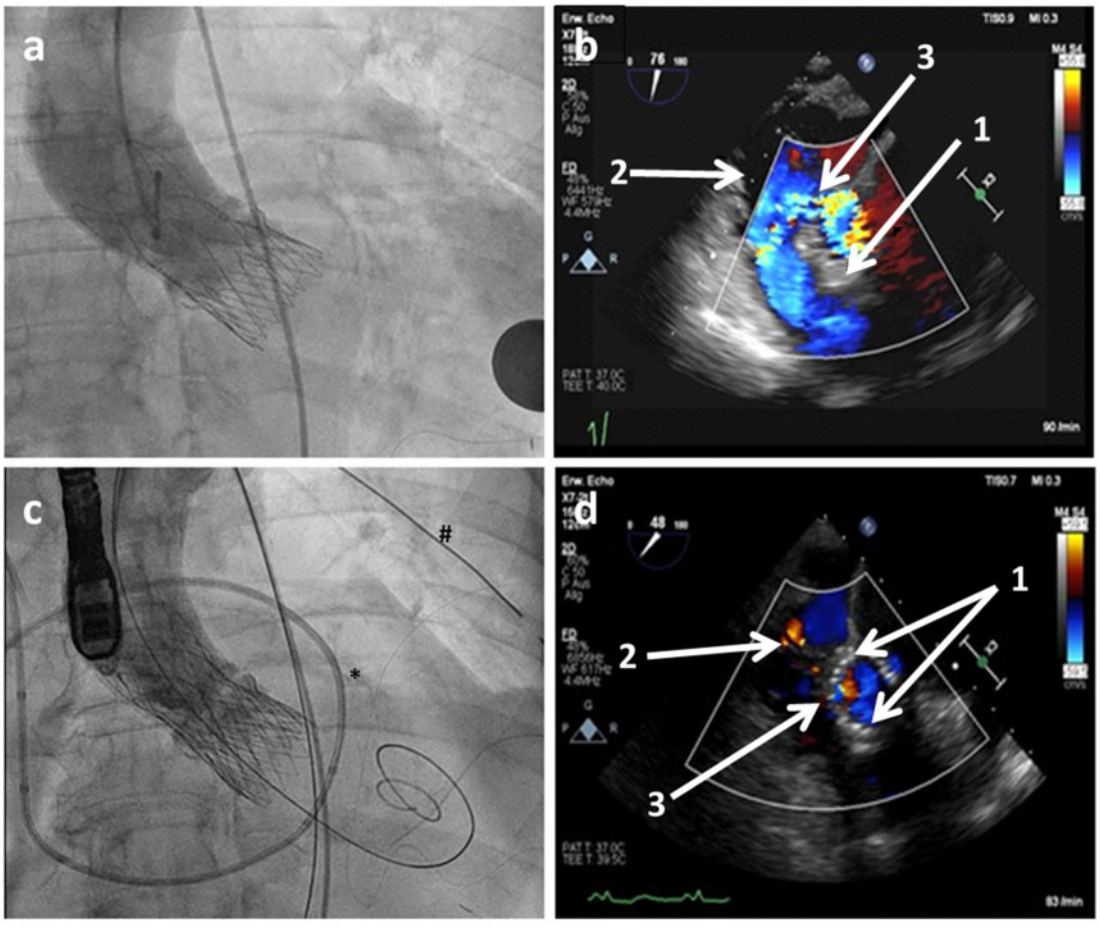

Figure 3 (*A*) Fluoroscopic image after transapical transcatheter aortic valve replacement (ACURATE neo M); (*B*) transoesophageal echocardiography following transapical transcatheter aortic valve replacement showing a severe ventricular septal defect; (*C*) angiography after valve-in-valve implantation. The implantation depth of the second valve (EVOLUT Pro 29 mm) was slightly deeper in the left ventricular outflow tract; and (*D*) transoesophageal echocardiography after the valve-in-valve procedure showing a small residual shunt. (**1**) Stentstruts, (**2**) tricuspid valve, and (**3**) leakage (ventricular septal defect). *****Pulmonary artery catheter, #Pleural drain.

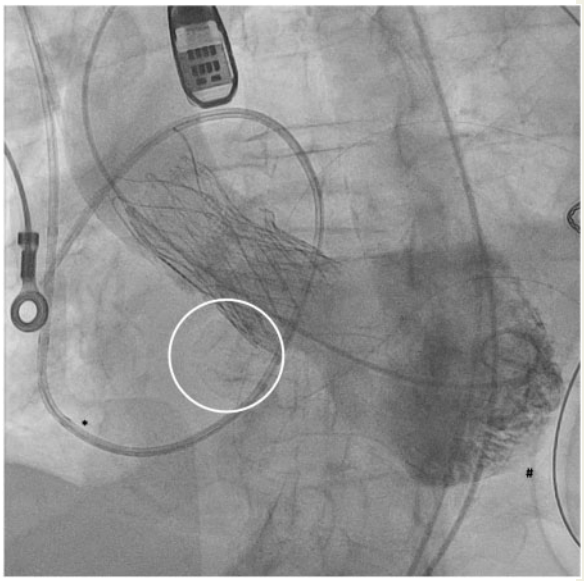

Figure 4 Left ventricular angiogram after valve-in-valve implantation showing a very small residual contrast shunt from the left-to-right ventricle (encircled). *Pulmonary artery catheter, #Pleural drain.

struts of the ACURATE valve and close to the aortic annulus (*Figure 3B*).

These findings were discussed and different therapeutic options were evaluated. Surgical treatment was rejected, and conservative management would lead to heart insufficiency due to the significant shunt. Implantation of an atrial or ventricular septal occluder might have disturbed valve function or generated an obstruction of the left ventricular outflow tract (LVOT). Implantation of a balloon-expandable valve was considered. However, there were concerns that it could worsen the situation due to the forces entering the annulus via the balloon. Implantation of a self-expanding valve was another option. We decided to perform a valve-in-valve (VIV) procedure via a transfemoral implantation of an EVOLUT Pro 29 mm (Medtronic Inc., Minneapolis, MN, USA), regardless of the elevated risk of vascular complications within the iliac arteries. This valve cannot, however, be implanted transapically. As subclavian arteries showed heavily calcified stenosis in the CT scan, transfemoral implantation was the only possible access route.

The VIV was implanted 2–3 mm below the lower edge of the first valve and towards the LVOT (*Figure 3C*). The implantation procedure remained uneventful and VSD was reduced to a small residual shunt without haemodynamic relevance (*Figures 3D* and 4). Furthermore, the PVL, which had been observed after first implantation, disappeared. Unfortunately, a pacemaker had to be implanted due to a high-grade AV block. On echocardiogram, valve functioning was normal, with a mean gradient of 8 mmHg. There were no signs of patient prosthesis mismatch (indexed effective opening area 1.0 cm^2, according to the continuity equation). A telephone-based follow-up 20 weeks post-procedure revealed complete recovery from the TAVR and improvement of the heart failure symptoms to NYHA class II. A follow-up echocardiogram was obtained 12 weeks following valve implantation and showed a persistently good result. A small VSD remained verifiable, but no PVL, and the mean gradient across the aortic valve was 8 mmHg. According to the 2020 European Society of Cardiology guidelines for the management of adult congenital heart disease, patients with a small residual VSD should be seen after 3–5 years. We recommended another echocardiogram one year following implantation.

Discussion

There are few case reports that describe iatrogenic VSD following TAVR. Most patients are treated conservatively or with an atrial septal defect[5–9] or VSD occluder.[10] In a systematic review, Ando *et al.*[11] identified 18 case reports that included 20 patients. There was only one case that described a VSD treated with a VIV procedure,[6] which fixed the VSD post-TAVR with a balloon-expandable valve. Ours is the first VSD to be fixed using the VIV-implantation of a self-expanding prosthesis, and we suggest this should be considered a feasible bail-out method for treating iatrogenic VSDs located near the annulus. The main leakage we observed was due to the VSD and was not paravalvular. Therefore, the valve must be implanted slightly more towards the LVOT than it is usual. Special care must also be taken to ensure the mitral leaflets are not affected by the implanted device. This was checked intra-procedurally via TOE in the long-axis view. The motion of the anterior mitral leaflet was not affected by the aortic prosthesis and no mitral insufficiency occurred. Given the supra-annular valve design, the EVOLUT Pro valve can be implanted slightly more towards the LVOT with an annular position of the new valve. However, this increases the risk of atrioventricular conduction-delays and the need for a pacemaker, as in our case, because of the well-known association between valve position and atrioventricular conduction delay. Hence, the deeper the valve is implanted into the LVOT, the higher the risk of an AV block.[12]

Balloon expansions are described as a potential reason for annulus ruptures. In our patient, the rupture was most likely caused by post-dilation. This highlights the advantage of a self-expanding valve in regard to the risk of annulus rupture. Some valves have an outer skirt of tissue that prevents PVL. In our case, it seals the leakage into the right ventricle. The ACURATE neo valve has a serrated ventricular edge, while the EVOLUT Pro has a smoother edge (*Figure 5A* and *B*). The leakage we observed was located between the struts of the serrated edge of the ACURATE neo valve (*Figure 5C*), therefore, we chose to implant an EVOLUT Pro as a second valve (VIV) (*Figure 5D*).

Patient perspective

While all of the complications led to a prolonged hospitalization stay, the patient was able to leave the hospital in good clinical condition.

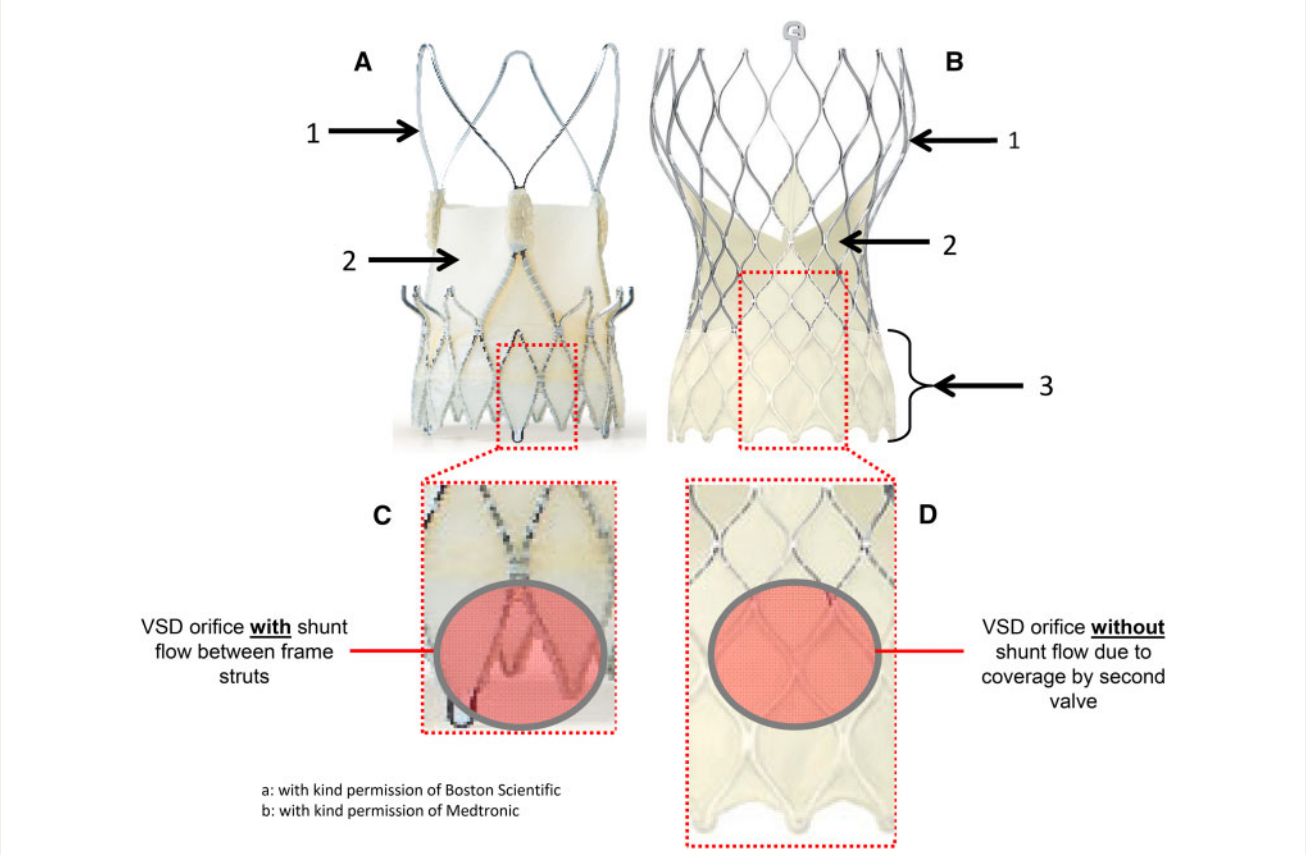

Figure 5 (*A*) ACURATE neo aortic valve; (*B*) EVOLUT Pro aortic valve; (*C*) ventricular septal defect projected onto the ACURATE neo valve; (*D*) ventricular septal defect projected onto the EVOLUT Pro aortic valve that was implanted into the ACURATE neo valve (valve-in-valve) and covered the defect. (**1**) Frame, (**2**) valve-leaflets, and (**3**) outer skirt.

During a telephone follow-up 4-month post-procedure, the patient reported that she was able to return to normal activities without serious health problems (NYHA II).

Lead author biography

Klaus-Dieter Hönemann, MD, is an interventional cardiologist. He graduated from University of Göttingen, Germany and received his doctorate in 1998. Since 2011, he is working at the Centre for Cardiovascular Disease, Schuechtermann-Klinik, Bad Rothenfelde, Germany. His clinical focus is structural heart disease and coronary heart disease. The Schuechtermann Klinik is a high-volume centre for interventional valve therapies with approximately 700 TAVR procedures annually.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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