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CASE REPORT
Cardiac imaging

EchoNavigator® technology facilitates transapical mitral paravalvular leak closure: a case report

Igor Vyacheslavovich Buzaev [1,2], Gulchachak Khalikova [1], Vladimir Vyacheslavovich Plechev [1,2], and Eustaquio Maria Onorato [2,3]*

[1]Republican Heart Centre, Ufa, Russia; [2]Bashkir State Medical University, Scientific Center of the Russian Academy of Science, Ufa, Russia; and [3]Centro Cardiologico Monzino, IRCCS University School of Milan, Via C. Parea, 4, 20138 Milan, Italy

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Background Catheter-based closure has emerged as a less invasive alternative to surgery in high-risk patients with paravalvular leak (PVL) and clinically significant regurgitation with feasibility and efficacy demonstrated in multiple studies.

Case summary A 72-year-old female with a past history of long-standing rheumatic heart disease underwent mechanical mitral valve replacement in 2008. Ten years later, redo surgery was performed due to a worsening mitral PVL and the leakage was closed by direct pledget-supported sutures, preserving the mechanical valve. She was recently admitted again for haemolytic anaemia and congestive heart failure (New York Heart Association Classes III–IV) due to a recurrent mitral PVL. We report our initial clinical experience using a novel software solution (EchoNavigator®-system) for intuitive guidance during a catheter-based transapical mitral PVL closure.

Discussion Transapical mitral PVL closure with a specifically designed device demonstrated in our case to be a better option than redo surgery. Recently introduced fusion imaging modalities enhanced visualization of soft tissue anatomy and device location improving enormously the results of this challenging intervention.

Keywords Fusion imaging modalities • Intraprocedural guidance • Percutaneous interventions • Paravalvular leak • Echocardiography • Case report

Learning points

- For high-risk symptomatic patients, catheter-based closure of paravalvular leak with a specifically designed device is a viable alternative strategy to surgical repair and may often represent the only available therapeutic option.
- Intraprocedural fusion of real-time three-dimensional transoesophageal echocardiography and cardiac fluoroscopy imaging using EchoNavigator®-system on the same display in real-time has a potential to improve the navigation, safety, and efficacy of this technically challenging procedure.

* Corresponding author. Tel: +39 348 6939883, Email: eustaquio.onorato@gmail.com
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Introduction

Based on the 2017 ESC guidelines, transcatheter closure may be considered for paravalvular leak (PVL) with clinically significant regurgitation in surgical high-risk patients (Heart Team decision) (Class IIb, level of evidence C).[1] Outcomes of transcatheter PVL closures have significantly improved over the years with growing experience and pre-procedural planning using multimodality imaging tools.[2,3] Current intraprocedural imaging techniques have several limitations: while fluoroscopy gives poor visualization of cardiac anatomical structures, echocardiography is limited by echo shadowing in its ability to detect the position of the catheters and devices. We report our initial experience with a novel software technology, the EchoNavigator®-system (Philips Healthcare, Best, The Netherlands).[4]

Timeline

Day 1	Patient admitted for heart failure and haemolytic anaemia
Day 2	Chest X-ray revealed cardiomegaly with increased enlargement of the pulmonary vasculature and central pulmonary artery dilatation. Electrocardiogram showed atrial fibrillation
Day 3	Two-dimensional transthoracic echocardiography (TTE) colour Doppler showed significant right ventricular volume overload, tricuspid annular plane systolic excursion of 17 mm, severe biatrial dilation, estimated systolic pulmonary artery pressure of 75 mmHg, dilated left ventricle with an ejection fraction of 60%
Day 4	Transoesophageal echocardiography (TOE) colour Doppler showed a crescent-shaped, 22 mm × 10 mm, posteriorly located (6 o'clock) mitral paraprosthetic leak with severe regurgitation
Day 5	The patient underwent a successful catheter-based closure of her recurrent mitral paravalvular leak with an 18 mm × 10 mm rectangular waist paravalvular leak device in a hybrid operating room under general anaesthesia and fusion of real-time three-dimensional TOE and cardiac fluoroscopy imaging using EchoNavigator®-system
Day 10	TTE at discharge confirmed the stable position of the device with trivial residual leakage. Clinical conditions greatly improved
Day 60	TTE confirmed the abolition of residual leak and normal function of the mechanical mitral prosthesis. Persistent clinical improvement with no need for further blood transfusions

Case presentation

A 72-year-old female was admitted for haemolytic anaemia, congestive heart failure (New York Heart Association Classes III–IV), and chronic kidney disease. The patient's medical history was notable for long-standing rheumatic heart disease, arterial hypertension, and permanent atrial fibrillation. Bileaflet mechanical mitral valve replacement (LIKS-28), tricuspid valve, and left atrium (LA) plasty were accomplished in 2008. Ten years later, due to a worsening mitral PVL, a thorough clinical and echocardiographic evaluation was performed and the suspicion of infective endocarditis was ruled out by negative blood culture tests. Redo surgery was performed and the leakage was closed by direct pledget-supported sutures, preserving the mechanical valve. On admission, bilateral basal crepitations, elevated jugular venous pulse, and mild lower limb oedema were found. Electrocardiogram showed atrial fibrillation with an average heart rate of 86 b.p.m. and diffuse ST-segment abnormalities. Chest X-ray revealed signs of widened cardiothoracic ratio, interstitial oedema, and pulmonary venous blood diversion. The patient was on diuretics, aldosterone inhibitors, betablockers, anticoagulants (warfarin), angiotensin-converting enzyme inhibitors, and amiodarone. She received occasional blood transfusions. Laboratory test results showed an increase of brain natriuretic peptide (452 pg/mL; normal values <125 pg/mL), serum lactate dehydrogenase (896 units/L; normal values: 140–280 units/L), and reticulocyte count (4.6%; normal values: 0.5–2.5%) and decreased levels of haptoglobin (30 mg/dL; normal values: 50–220 mg/dL) and haemoglobin (10 g/dL; normal values for women: 12.0–15.5 g/dL). Two-dimensional (2D) transthoracic echocardiography (TTE) colour Doppler showed severe biatrial dilation, significantly enlarged right ventricle with a tricuspid annular plane systolic excursion of 17 mm, estimated systolic pulmonary artery pressure of 75 mmHg, and dilated left ventricle (LV) with an ejection fraction of 60%. Three-dimensional (3D) transoesophageal echocardiography (TOE) colour Doppler showed no prosthetic dysfunction, a huge crescent-shaped posteriorly located (6–7 o'clock) mitral PVL (*Figure 1*) with severe regurgitation (PISA 1.2 cm, vena contracta 0.9 cm, regurgitant volume 90 mL, EROA 1.2 cm^2). By cropping of regurgitant jet volume on 3D colour Doppler TOE image the size of the leak was measured to be 22 mm × 10 mm in diameter. Coronary angiography showed a stenosis of 40% on the first obtuse marginal branch of the left circumflex artery.

After heart team discussion and due to prohibitive surgical risk, it was decided to address her recurrent mitral PVL using a catheter-based technique. The patient was consented after the local ethics committee and the local regulatory agency have approved the use of the device. The procedure was carried out in a hybrid operating room under general anaesthesia, continuous real-time 2D/3D TOE colour flow Doppler, and fluoroscopic guidance with the presence of both interventional and surgical teams. Left anterolateral thoracotomy was performed and the LV apex was visualized. Pledgeted purse-string sutures were then placed at the apex and an access needle was introduced to puncture the apex. Once in the LV cavity, a 0.035″ standard guidewire was introduced via the needle and the needle was exchanged for a short 7 Fr sheath.

Fusion of real-time 3D TOE and cardiac fluoroscopy imaging was obtained using EchoNavigator®-system. The location of the PVL was marked to aid the pathway of the guidewire crossing

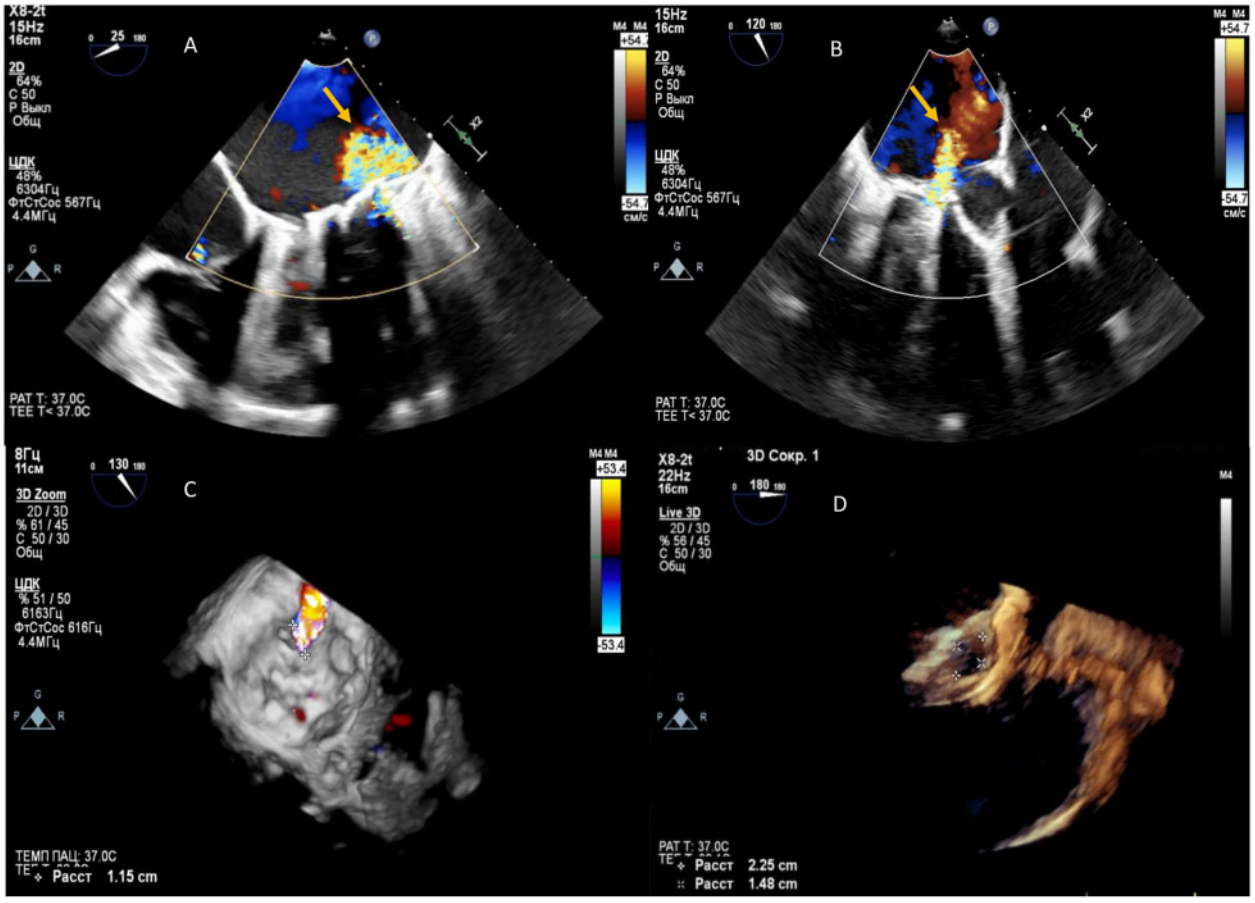
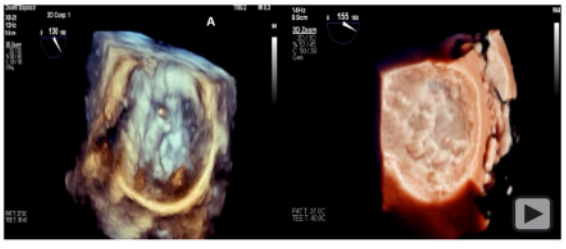


Figure 1 Two-dimensional transoesophageal echocardiography colour flow Doppler at 25° and 120° views showing the partially eccentric significant regurgitant paravalvular leak jet (orange arrow) (*A* and *B*) and three-dimensional transoesophageal echocardiography images showing leakage measurements (*C* and *D*).

the PVL. Through the short 7 Fr sheath at LV apex, a 0.035″–180 cm highly hydrophilic Glidewire® (GR3508, Terumo) was used to get across the leakage into the LA (*Video 1*) with the help of a 6 Fr multipurpose C guiding catheter. The guiding catheter was advanced into the LA and the hydrophilic wire replaced with a 0.035″–260 cm Amplatz extra-stiff wire (*Figure 2*). Over the extra-stiff wire placed in the LA, a 12 Fr–28 cm in length Sentrant introducer sheath with hydrophilic coating (Medtronic) was advanced crossing the leakage: through the sheath an 18 mm × 10 mm rectangular waist paravalvular leak device (PLD, Occlutech, Helsingborg, Sweden) was advanced into the LA (*Figure 3* and Supplementary material online, *Figure S1*). The self-expanding, flexible, double-disc device made from nitinol-braided wires (LA disc, 28.5 mm; LV disc, 26.5 mm) was deployed under fusion imaging modalities guidance. A correct device alignment to the defect with significant decrease of the regurgitant jet was achieved without impingement on the prosthetic discs (*Figure 4* and *Video 2*). A pull-and-push manoeuver was done to check the PLD stability and the device was finally released. Transillumination rendering 3D TOE (TrueVue, Philips Medical Systems) images nicely showed the final position and orientation of the device (*Figure 5* and *Video 3*). The access site



Video 1 Real-time three-dimensional transoesophageal echocardiography (*A*) and transillumination rendering three-dimensional transoesophageal echocardiography (TrueVue, Philips Medical Systems) (*B*) videos showing the retrograde guidewire advancement into the left atrium across the mitral paravalvular leak.

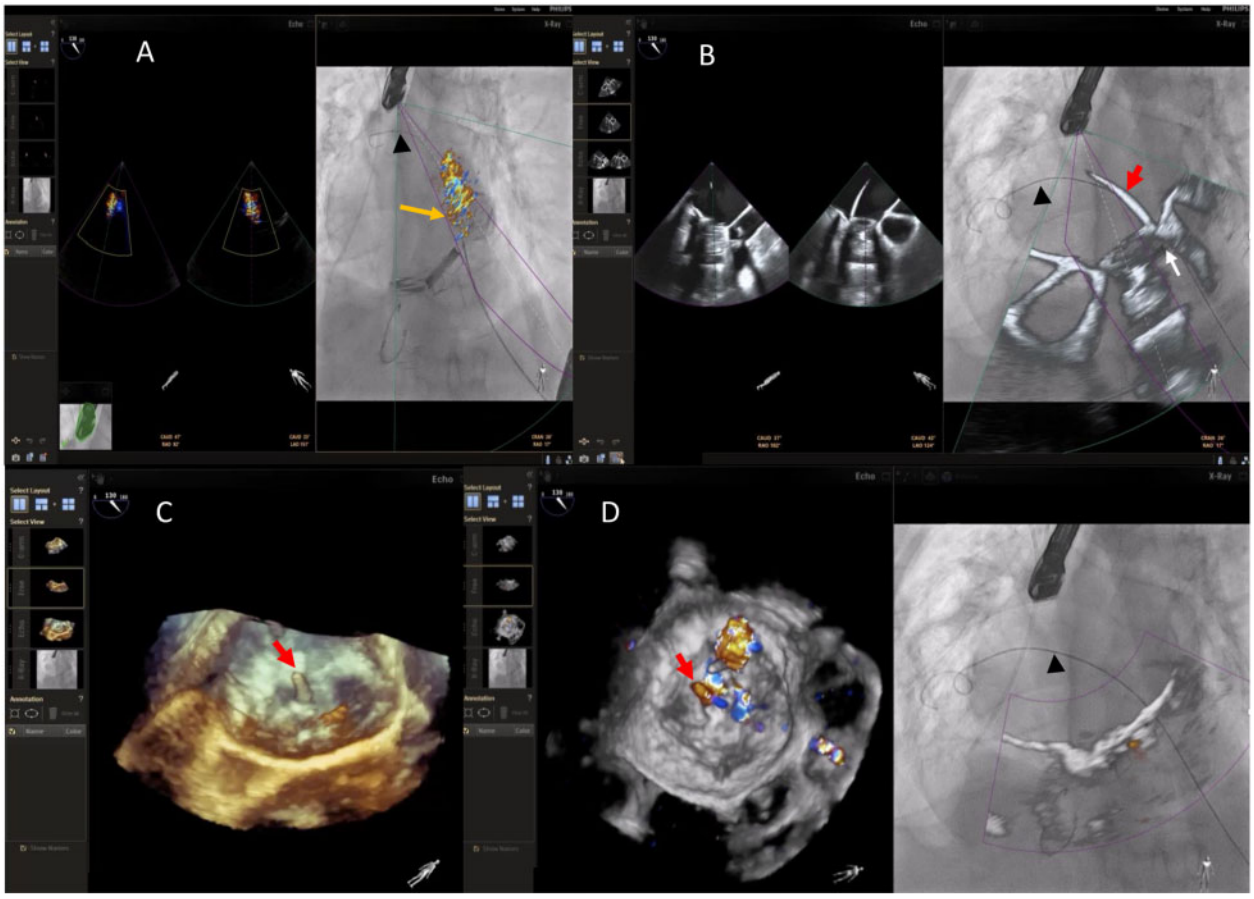


Figure 2 Intraprocedural fusion of fluoroscopy and real-time two-dimensional/three-dimensional transoesophageal echocardiography colour flow Doppler at 130° view showing the regurgitant paravalvular leak jet (orange arrow) (*A*) and the guiding catheter (red arrow) passage through the leakage (white arrow) with the fused images maintained (stiff wire in the left atrium, black arrowhead) in two-dimensional X-plain views (*B*); real-time three-dimensional transoesophageal echocardiography colour flow Doppler images showing the tip of the guiding catheter (red arrowhead) (*C*) and regurgitant jet around it (*D*).

was closed surgically at the completion of the procedure. The postoperative course was uneventful and the patient was discharged home on the fifth postoperative day without complications. TTE confirmed stable position of the device with trivial residual leak.

At 2-month follow-up the patient had a significant clinical improvement with no need for further blood transfusions. TTE confirmed the abolition of residual leak, normal function of the mechanical mitral prosthesis, and an estimated pulmonary artery pressure of 45 mmHg.

Discussion

EchoNavigator®-system allows unique real-time fusion of live X-ray and live echo images for intuitive guidance during mitral PVL closure procedure providing live dynamic imaging and allowing the interventionalist to view catheters and devices (by fluoroscopy) and soft tissue anatomy (by echocardiography) in the same image and orientation. The software requires a full echocardiographic and fluoroscopic Philips system. The ultrasound system must include the 3D modality, using a standard TOE probe (X7-2t; Philips Healthcare) combined with a recent fluoroscopic system (Allura Xper FD10; Philips Healthcare). This real-time and user-friendly software requires no additional hardware to merge and synchronize the echocardiographic and fluoroscopic modalities. The addition of an annotation marker at the site of the paravalvular defect may be helpful, particularly in cases where the defect is small and not well seen by 3D echocardiography. The incremental value of fusion modalities is key in cases of fluoroscopically invisible mitral bioprosthesis or anatomically hard-to-approach calcified serpiginous tracts. Nevertheless, the adoption of this technology has not been widespread for some reasons: first, the addition of fusion modalities is not always necessary, particularly when structures of

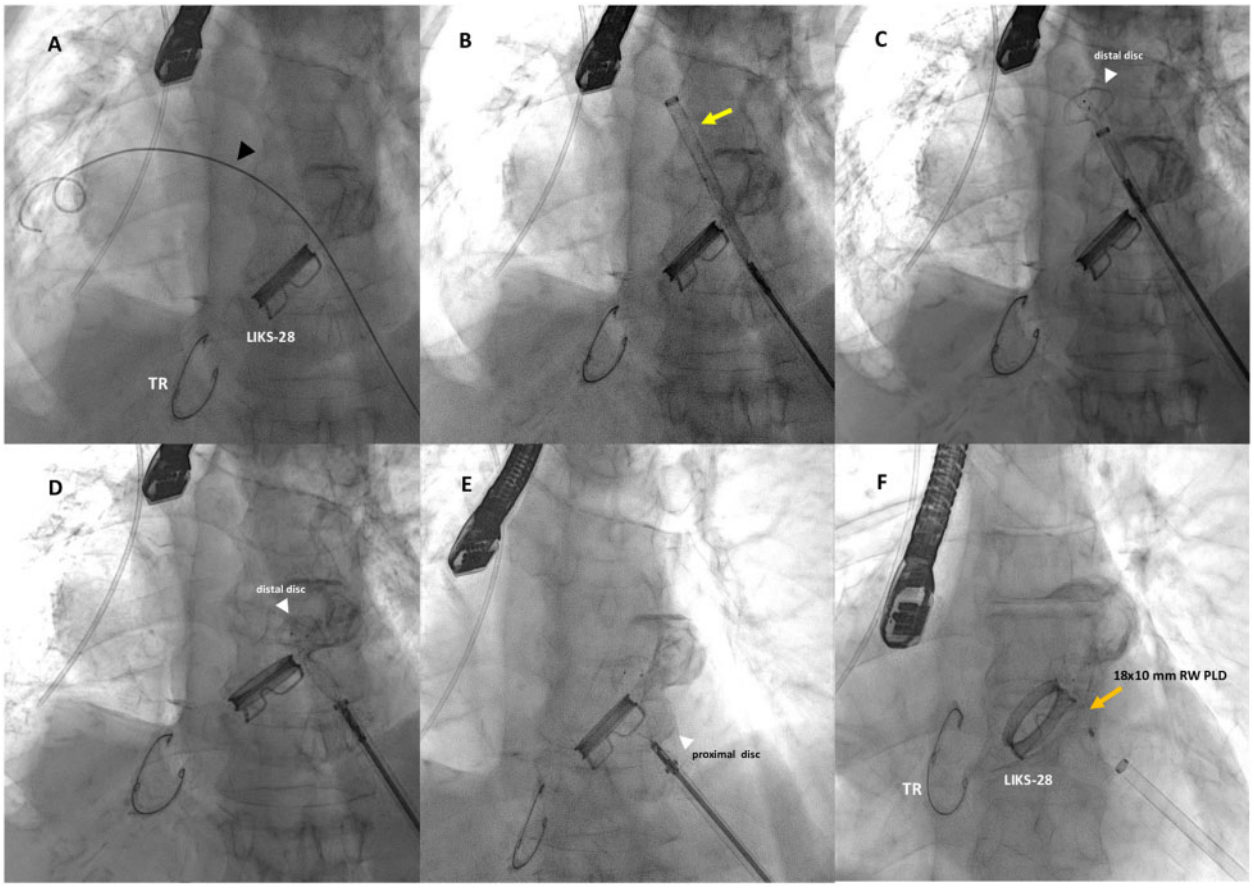


Figure 3 Fluoro-angiographic procedural steps. (*A*) From the transapical approach an extra-stiff wire (black arrowhead) across the leakage with the distal soft tip in the left atrium; (*B* and *C*) the distal part of a 10 Fr delivery system (yellow arrow) in the left atrium and the distal disc opening (white arrowhead); (*D* and *E*) distal and proximal discs (white arrowheads) of the 18 mm × 10 mm rectangular waist paravalvular leak device positioning; (*F*) the paravalvular leak device (orange arrow) finally deployed. LIKS-28, bileaflet mechanical mitral valve; TR, tricuspid ring.

interest are seen well by a single imaging modality; second, co-registration of the TOE probe with the gantry might even lead to a temporary increase in radiation exposure.[5] Prospective randomized multi-centre studies with a larger sample size are necessary to demonstrate the real benefits of this promising technology.

No randomized studies have compared surgery with transcatheter techniques in terms of PVL management.[6,7] Redo surgery for mitral paravalvular defects may definitively carry high morbidity, mortality, and high rate of reoccurrence.[8] It is very likely that the percutaneous approach should not be limited to critically ill patients rejected for surgery but should be considered the therapeutic option of choice in experienced centres.[9]

Mitral PVL can be approached via antegrade trans-septal access particularly suitable for antero-laterally located leaks or via retrograde transaortic approach from the LV. An open surgical retrograde transapical access (hybrid technique) was conversely chosen in our case. Important advantages of the hybrid approach are (i) coaxiality to the mitral plane, direct engagement to the leak, and crossing on the same direction of the regurgitant flow; (ii) better wire pushability and easier access to leakages particularly for posteroseptal ones; and (iii) controlled surgical closure of the ventricular puncture.[10,11] The disadvantage is represented by a more invasive approach. Further studies comparing different access sites for percutaneous PVL closure are needed.

The frequently used occluders in the early experience have been created for other applications, such as atrial/ventricular septal defect or patent ductus arteriosus closure. The Amplatzer Vascular Plugs (AVP), in particular the oblong AVP III, have a clear advantage over other occluders with a circular cross-section. Nevertheless, the major disadvantage of this otherwise applicable device is the lack of fabric inside the nitinol mesh that forces operators to implant multiple plugs to achieve dense filling of the PVL channel.[12]

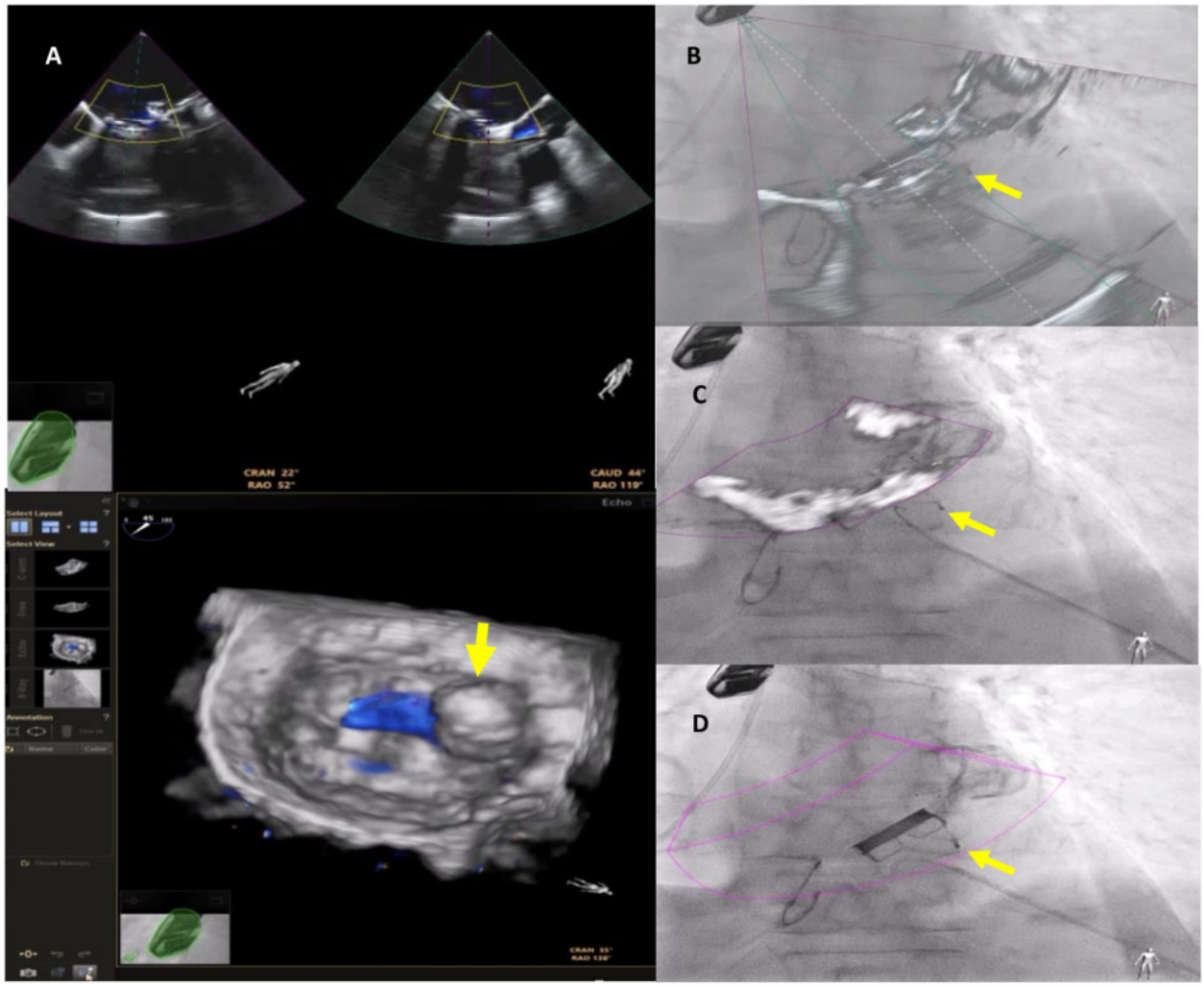
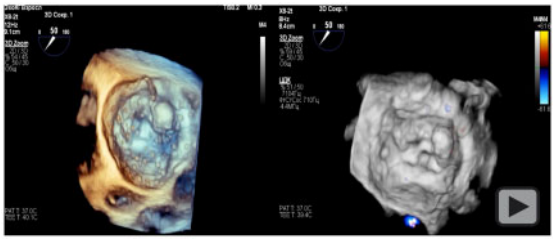


Figure 4 Fusion of real-time two-dimensional/three-dimensional transoesophageal echocardiography colour flow Doppler at 45° (*A*) and fluoroscopic images (*B–D*) with the fused images maintained demonstrating the correct position of the paravalvular leak device (yellow arrow) without impingement on the prosthetic discs or surrounding structures.



Video 2 Real-time three-dimensional transoesophageal echocardiography colour flow Doppler videos demonstrating the correct position of the paravalvular leak device.

A specifically designed CE-marked device like PLD, available in 4 different shapes and 19 different sizes, might allow PVL closure to be performed more conveniently and efficaciously, mostly often with one single occluder.[13,14,15]

Conclusion

EchoNavigator®-system has the potential to increase the safety, accuracy, and effectiveness of catheter-based PVL closure procedures by enhancing the understanding of anatomical structures and the spatial relation between the X-ray and ultrasound images. However, further studies are needed to assess the added value of this new technology.

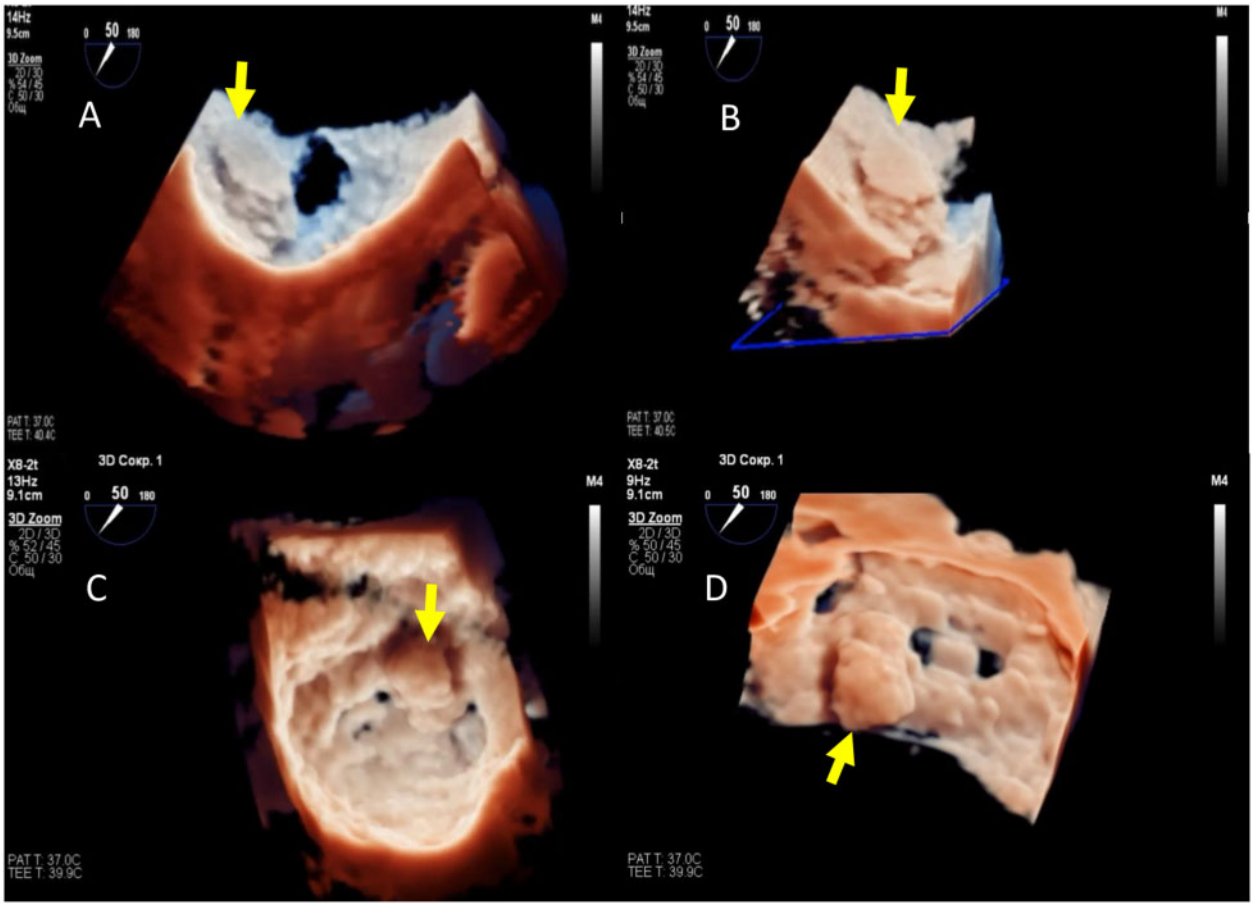
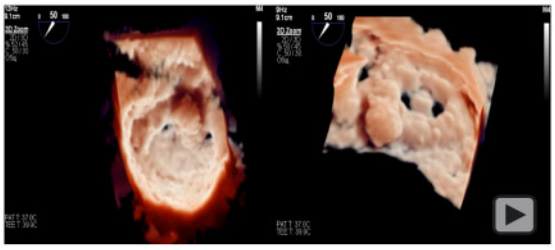


Figure 5 Transillumination rendering three-dimensional transoesophageal echocardiography (TrueVue, Philips Medical Systems) images (*A–D*) showing the final position and orientation of the occluder device (yellow arrow).



Video 3 Transillumination rendering three-dimensional transoesophageal echocardiography (TrueVue, Philips Medical Systems) videos showing the appropriate position of the 18 mm × 10 mm paravalvular leak device, its relationship with the surrounding structures (*A*) and the correct functionality of the mechanical prosthetic valve (*B*).

Lead author biography

Dr Igor Vyacheslavovich Buzaev, MD, PhD, is an interventional adult and paediatric cardiologist, particularly active and skilled in coronary and structural congenital heart disease interventions at Republic Heart Centre in Ufa, Russia. He is a Professor and Surgery Chair at Bashkir State Medical University, Ufa and Senior Consultant of Bashkortostan Republic Healthcare Ministry in Interventional Cardiology. He has been awarded as Advanced Doctor of Medicine degree (DSc) by the Ministry of Science and Higher Education of the Russian Federation (2019). He is the co-

founder of the first Russian Neuro-MRGFUS site at International Medical Centre 'V.S. Buzaev memorial' in Ufa (2020). He is the current Chief of Interventional Department in Republican Heart Centre, Ufa, Russia.

Supplementary material

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

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

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: Eustaquio Maria Onorato is a consultant for Occlutech, manufacturer of the device. The remaining Authors declare no commercial or financial relationships that could be construed as a potential conflict of interest.

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