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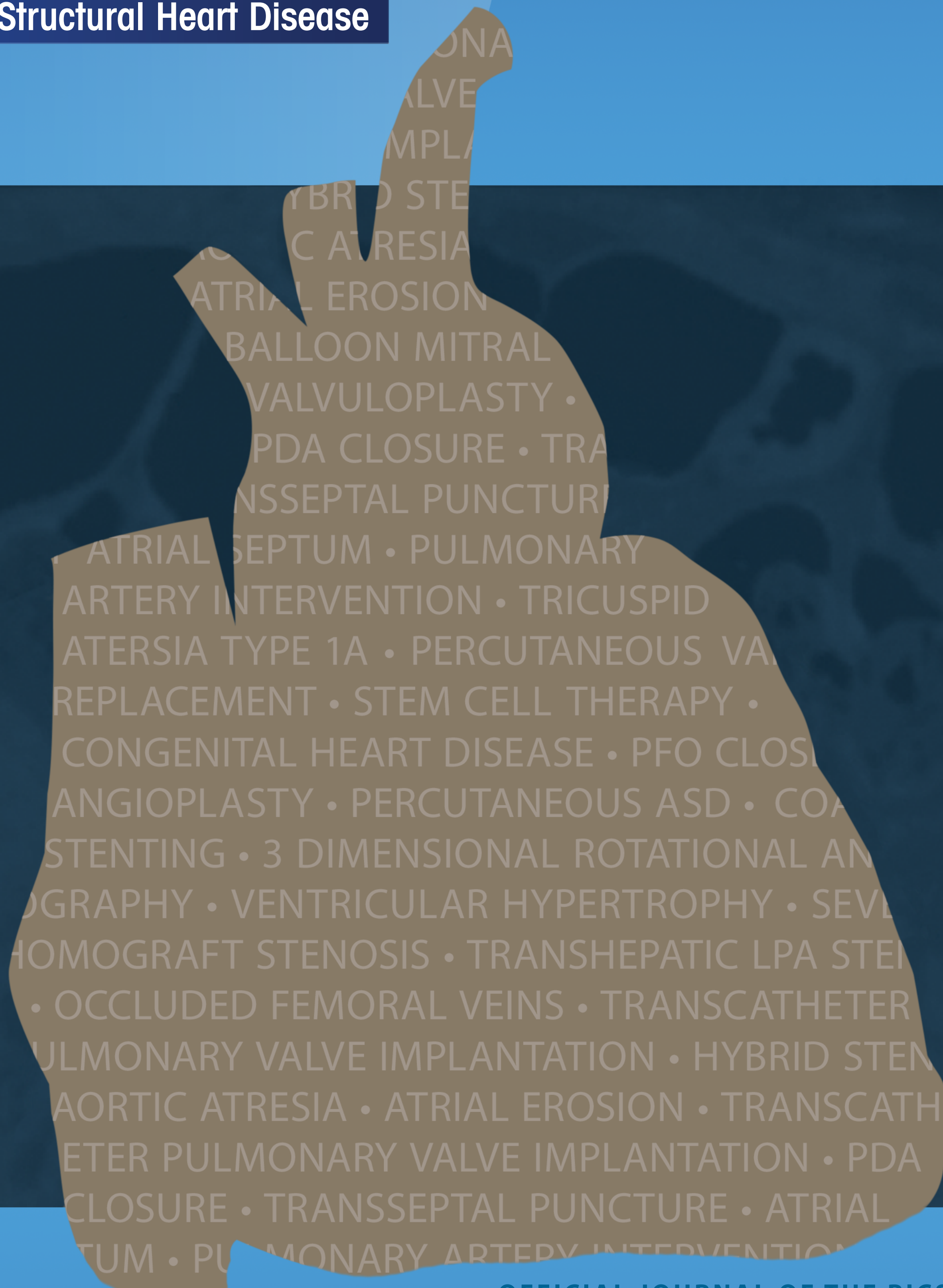

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Transcatheter Mitral Valve Replacement: Update on New Devices

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Abstract

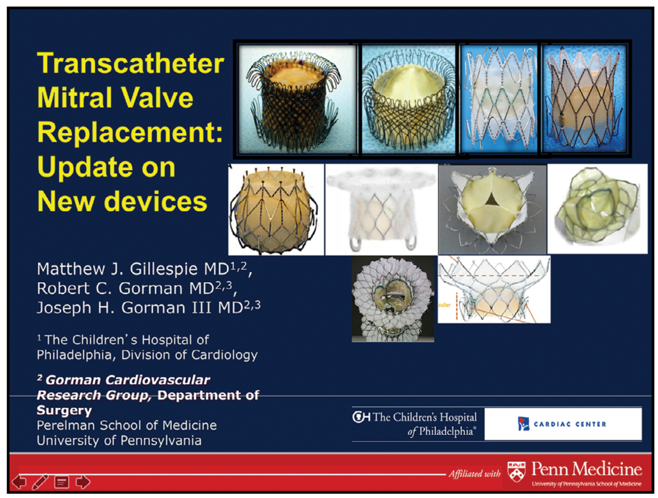
Transcatheter Mitral valve replacement represents the next frontier in cardiac valve therapy. This review article details the authors' experience thus far in the development of a novel catheter-based mitral valve replacement device, and also highlights the preliminary experiences of other new technologies.

Key Words

Mitral valve replacement • Mitral regurgitation • Transcatheter valve replacement • Ischemic mitral regurgitation • General

Conflict of Interest

The authors each have ownership interest in Annulon LLC, which is a company formed around this and other mitral valve technologies



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Atrial Septal Defects: Special Considerations in Management and Closure in the Adult Patient Population

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Abstract

Atrial septal defects are the third most common type of congenital heart disease, and include several types of atrial communications that allow shunting of blood between the systemic and the pulmonary circulations. Common manifestations in adult patients with untreated defects include exercise intolerance and atrial tachyarrhythmias. If remained untreated, right ventricular dysfunction, and pulmonary hypertension may ensue, leading to a lower quality of life and premature death. Treatment options include surgical and percutaneous transcatheter closure. Surgical closure is safe and effective and when done before age 25 years is associated with normal life expectancy. Transcatheter closure offers a less invasive alternative for patients with a secundum defect. In this review we discuss the anatomy, pathophysiology, treatment options with emphasis on technical aspects, and outcomes of atrial septal defects in adult patients. We provide in depth discussion on the clinical scenarios that are unique to this patient population, including septal defects in the constellation of pulmonary hypertension and diastolic dysfunction.

Key Words

Atrial septal defect • Adult congenital

Conflict of Interest

The authors have no conflict of interest relevant to this publication.

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Coronary Artery Fistulae: Indications For Treatment and Technical Considerations

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Abstract

Coronary artery fistulae is an uncommon cardiac condition. Trans catheter closure of fistula remains the treatment of choice. Due to the condition being very uncommon, individual experience remains limited. In the present article, we have described various aspects of coronary artery fistula and their management.

Key Words

Coronary artery fistula • Transcatheter closure.

Introduction

Coronary artery fistulas (CAFs) are an uncommon cardiovascular anomaly characterized by a connection between a coronary artery and an adjacent vessel (coronary arteriovenous fistula) or a cardiac chamber (coronary-cameral fistula) without an intervening capillary network [1]. It is the result of a defect early in the development of the myocardium, prior to compaction, and leads to the persistence of sinusoids, resulting in a coronary-cameral fistula, whereas a defect at a later stage of development results in a CAF.

Incidence

The true incidence of CAFs is unknown, as the majority of patients remain clinically asymptomatic. Their presence may be suggested by an audible cardiac murmur, or by way of diagnostic imaging with either an echocardiogram or coronary angiography [2].

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The reported incidence is highly variable, ranging from 0.02 to 2.1%, depending on the modality of investigation [3]. In one series, an incidence of 2.1% was reported, but the majority of patients had left coronary artery to left ventricular micro-fistulae [4]. In a series of 126, 595 patients undergoing diagnostic coronary angiography, the incidence was 0.18%, and the majority were small and the patients were asymptomatic, suggesting that many of the CAFs were co-incidental findings [5]. In children, the incidence has been reported to be 0.06% from transthoracic echocardiography [6]. Interestingly, a review of literature between

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1994 and 2003 found only 236 pediatric and adult patients with CAFs [3]. These lesions can be associated with congenital heart disease, mainly with tetralogy of Fallot or pulmonary atresia [7, 8].

Site of Origin and Drainage

CAFs can arise anywhere in the coronary circulation, but nearly half come from the right coronary artery (RCA) (~51%), followed by the left coronary artery and its branches (~43%). In a small proportion of patients they originate from both coronaries (~5%). In the series reviewed here, nearly 90% of fistulas drained into the systemic venous circulation; the right ventricle (RV) (~41%), right atrium (RA) (~26%), pulmonary artery (PA) (~17%), coronary sinus (CS) (~7%), or superior vena cava (SVC) (~1%). CAFs have also been observed to drain into the left atrium (LA) (~5%) and left ventricle (LV) (~3%) [9]. In the majority of patients, the fistulas enter into their draining chamber/vessel through a single communicating channel, and a minority has multiple entry sites.

Etiologies and Types of CAFs

In the vast majority of patients, CAFs are present at birth, whereas trauma, myocardial biopsy, chest irradiation, and pacemaker implants can result in an acquired form. Angiographically, the fistulas can be described as proximal or distal, based upon their origin from the coronary artery. Proximal CAFs may be small or large, have single or multiple feeding vessels, originate from one or both coronaries, and are devoid of any nutritive branches. Distal fistulas are usually larger and have high flow [10]. There is no clear definition of what is considered a small, medium, or large fistula by measurement criteria. Generally, small fistulas are clinically silent and do not result in any long-term sequelae, whereas untreated large fistulas are more likely to be associated with myocardial ischemia/infarction, endocarditis, heart failure, rupture, and progressive dilation of the draining chamber/fistula [2].

Physiology of the CAF

Large CAFs result in (1) shunting of the blood from left to right side (in majority of patients), and (2) have the potential for a 'coronary-steal' phenomena, affecting blood flow to the myocardium supplied by the coronary artery distal to the origin of the fistula. These physiologic changes are influenced by the size of the fistula, pressure difference between the connecting chambers, and co-existence of coronary artery disease. In the only reported animal model (adult foxhounds) of a CAF, the circumflex (LCx) coronary artery was connected to the PA with a vein graft. Calculated mean flow through the fistula was 89 ml/min, resulting in Qp:Qs of 1.1:1. Increasing proximal coronary flow by 211% and producing a relative steal of 26% from the coronary artery distal to the fistulas [11]. Doppler wire assessment in a patient with a CAF demonstrated higher systolic and blunted diastolic flow-velocities, which normalized after successful closure [12].

Clinical Presentation & Differences Based upon Age Group

Analysis of data published between 1993 and 2004 on patients with CAF, demonstrated that 117/128 (91%) adult patients were symptomatic vs. 105/133 (79%) pediatric patients who were asymptomatic. Chest pain and dyspnea were the most common symptoms (71%), but patients also presented with palpitations (6.5%), fatigue (6.5%), pre-syncope/syncope (5.6%), congestive heart failure (3%), pulmonary hypertension, fistula thrombosis, and rarely rupture or infective endocarditis. An auscultatory finding of a murmur (continuous, systolic or diastolic) was a common physical sign (40%). The fistulas can grow in size over time and result in symptoms if left untreated. Spontaneous closure and infective endocarditis were more common in the pediatric age group, whereas aneurysmal dilation (14%), spontaneous rupture (4%), and co-existing coronary artery disease (19%) were seen mainly in adult patients [3].

Indication for Treatment and Risk Management

Our understanding of the natural history of patients with CAFs is significantly limited. According to the American College of Cardiology congenital guidelines, patients with large CAFs should be considered for surgical or transcatheter closure (TCC) irrespective of symptoms (Class I, level of evidence C). Additionally, patients with small to moderate fistula, in the presence of documented evidence of myocardial ischemia, arrhythmia, unexplained ventricular dilation or dysfunction, should also be treated with transcatheter or surgical closure [13]. Small fistulas can usually be left untreated and in some cases have been reported to close spontaneously [14]. Alternatively, patients with intermediate sized CAF can be followed with regular surveillance by echocardiogram or CT angiography, and treated if there is evidence of new-onset symptoms, ischemia or progressive dilation. Merely identification of a CAF does not necessitate treatment.

Treatment Options

The first successful surgical repair of a CAF was performed in 1956, and was the only treatment option until 1983, when TCC became a viable alternative [15, 16]. Surgical repair remains the treatment of choice for fistulas that are not suitable for TCC as well as in those undergoing surgical repair of other cardiac conditions, for example, congenital cardiac defect repair or coronary artery bypass surgery. Although surgical closure is a relatively low risk procedure, it is associated with inherent risk involved with the surgery. The TCC approach was first used in 1983 using a detachable balloon system [17], and over the years, various closure devices have become readily available, including coils, and vascular plugs. Choice of the device is based upon the size of the fistula, site of device delivery, required catheter size and length. Over the last three decades, TCC has become the preferred treatment option. The risks involved with the procedure include:

- Occluding coronary branches with the device,
- Embolization of clot from the occluded branch,
- Propagation of clot,
- Dissection of coronary artery due to catheter or device manipulation, and
- Arrhythmias

Technical Aspects of the CAF Closure

TCC of CAFs is a technically challenging procedure, and as the number of procedures performed is small, individual experience remains limited. The decision to treat a CAF should be based upon various factors including: the patient's age, symptoms, fistula size, possibility of progression or rupture, the origin and termination of the fistula, the number of feeding vessels, risk to viable myocardium, risk of residual vessel thrombosis, likelihood of success, and available resources including experience, devices, and personnel. Surgical consultation, specifically for large distal fistulas should almost always be obtained, especially if interventional experience is limited.

Utilization of other imaging modalities in addition to coronary angiography has proven useful for anatomical characterization and can aid strategizing the technical aspects of the procedure. Contrast-enhanced CT is a noninvasive and accurate imaging technique for the detection of CAFs. Multidetector CT has been shown to provide high-resolution anatomic images, and it allows evaluation of aneurysmal dilation or thrombus formation in the vessel. Volume-rendered images acquired from 3D CT data sets provide excellent overviews of the cardiac and vascular anatomy and can help the interventionist or surgeon understand the anatomical complexity.

An operator may choose either an antegrade (via venous circulation) or retrograde (via coronary circulation) approach. Retrograde approaches may be challenging due to anatomy, angulation, kinking, size of the delivery sheath required, and potential risk to coronaries vessels. TCC of proximal CAFs can be performed through a retrograde approach, whereas the same approach for closure of large and especially distal fistulas may be challenging. Delivering a device or coil into the distal part of the coronary circulation, which is likely to be tortuous, requires supportive catheters. Using a larger sized catheter or a catheter in a 'mother-child' format (telescoping) offers good support, but catheter length may be inadequate, especially in those with distal and tortuous lesions, or if the patient is very tall. In such cases antegrade approaches through the venous side would be preferred, although catheter engagement from the venous side can be challenging. Formation of an arterio-venous (AV) loop, where the fistula is wired from the coronary artery and then exteriorized on the venous side (with the help of snare) can be a useful strategy (cross one-way and close the other-way).

In the present article, we will review technical aspects of the TCC of CAFs by outlining various procedures we have performed over last few years.

Need for Larger Catheter

Blood flows at high velocity in large CAFs, and to opacify such vasculature, large volumes of contrast are required. Injecting through either 5 or 6 French (Fr.) sized catheter may not be adequate, and large caliber catheters may be required. As demonstrated here, a large CAF originating from the LCx artery is difficult to opacify with injection through 6 Fr. catheter (Figure 1A). Although an 8 Fr. catheter delineates the CAF better (Figure 1B), it failed to demonstrate the distal branches. The vessels were visualized only when distal part of the CAF was occluded with balloon wedge catheter inserted from the venous side over the AV loop (Figure 1C). Presence of a marker catheter in aorta may help with sizing the CAF (Figure 1D), if auto isocenter is not available on the angiographic equipment for calibration.

Retrograde Approach

A CAF originating from the proximal part of the coronary artery can be successfully treated through a retrograde approach. A 72-year-old man presented with chest pain and elevated troponin levels. An echocardiogram demonstrated inferior wall hypokinesia, and coronary angiography demonstrated no flow limiting coronary artery disease. There was a medium sized CAF originating from the RCA (Figure 2A & B). We treated this fistula using a retrograde approach with an Amplatz left (AL) 0.75-guide catheter and positioned a Renegrade® micro-catheter (Boston Scientific, Marlborough, Massachusetts, USA) over a Pilot® 50 guide wire into the fistula. We estimated the fistula diameter to be approximately 4 mm, so first of all we delivered a 5 mm × 15 cm IDC® platinum, interlock detachable coil (Boston Scientific Inc., USA). As there was residual flow, we delivered two further coils measuring 3 mm × 12 cm to close the fistula (Figure 2C & D). Adequate catheter support was very important, and

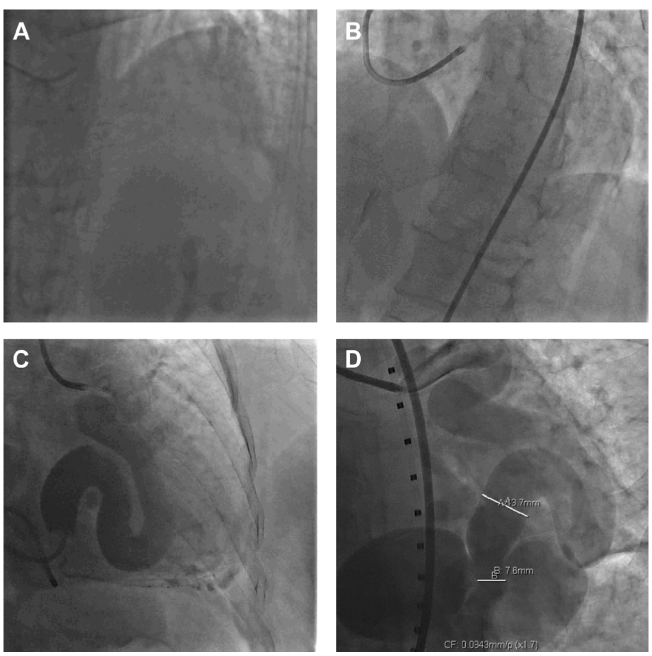

Figure 1. Large CAF – Large catheters. *Panel A.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.01. *Panel B.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.02. *Panel C.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.03. *Panel D.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.04.

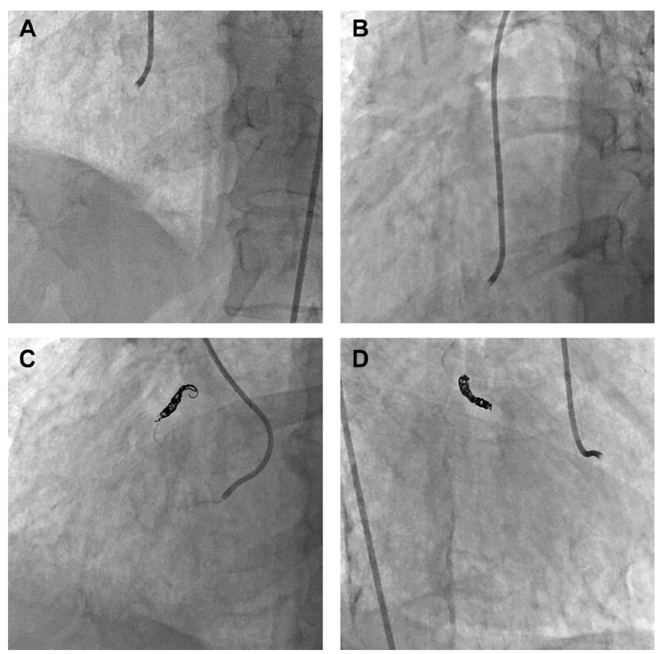

Figure 2. Retrograde approach. *Panel A.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.05. *Panel B.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.06. *Panel C.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.07. *Panel D.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.08.

as one can see here the catheter was pushed out of the RCA despite using an AL guide that are known to be very supportive. We initially chose coils that were nearly 25% larger in diameter than the CAF, followed by smaller diameter coils to fill up residual gaps until flow was compromised. In the present case, there was some residual sluggish flow at the end of the procedure that was very likely to stop completely over the next few days.

Antegrade Approach

A 54-year-old man presented with a symptomatic large CAF originating from the LCx draining into SVC. We used a 7 Fr. JL4 guide catheter to opacify the CAF. There was gross dilation of the left main and the proximal LCx. The left anterior descending (LAD) artery could not be well visualized due to the high flow state towards the LCx artery (Figure 3A). We used a system consisting of a soft glide exchange wire, a 4 Fr. GlideCath®, a 6 Fr. multipurpose coronary guide catheter, and the Agilis® sheath (St. Jude Medical Inc., USA) (Figure 3B & C). This allowed us to make progress with a 6 Fr. guide catheter to deliver a 12-mm Amplatz Vascular Plug® II, to successfully occlude the fistula (Figure 3D).

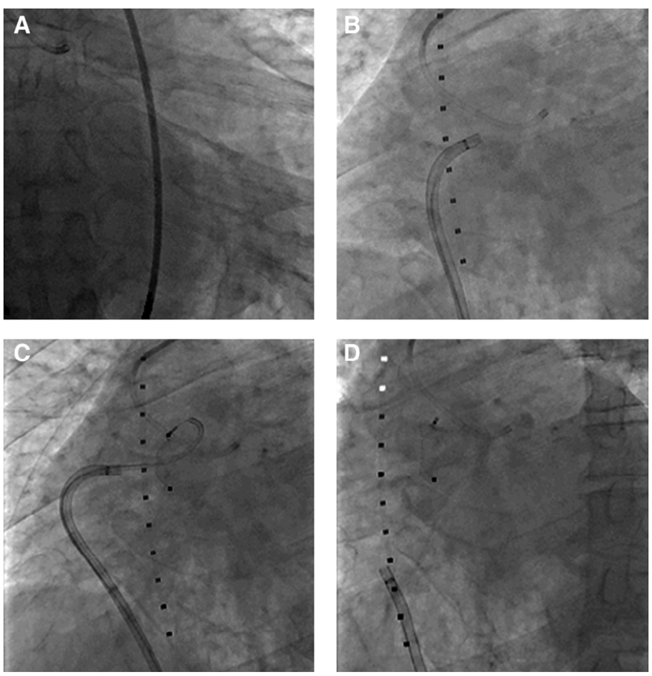

Figure 3. Antegrade closure. *Panel A.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.09. *Panel B.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.10. *Panel C.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.11. *Panel D.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.12

This case demonstrates that if the connection of the fistula on the venous side can be clearly observed, it is possible to perform the TCC from the venous side with help of supportive sheaths and catheters, without need for AV loop formation.

Arterio-Venous Loop Formation

TCC of large-sized distal CAFs requires closure device and/or coils to be delivered distally, which can be extremely challenging, and on occasion be better performed through an antegrade approach. Cannulation of the draining end of the CAF is challenging, due to difficulty in precise localization and angulation. An AV loop may assist overcoming these challenges. Figure 4A-D demonstrates a patient with a giant CAF originating from the LCx artery, draining into the CS (Figure 4A). The landing site for the device was very distal and we decided to deliver the device through an antegrade approach, over an AV loop. We used an 8 Fr. guide catheter to obtain adequate support and initially used 0.035-inch soft Glidewire® (Terumo Interventional System, Japan), which just looped but did not exit the CS (Figure 4B). We then used a 300-cm exchange length Prowater® guide wire supported by a Finecross® micro-catheter (Terumo Interventional System, Japan) to traverse through CS into the RA, where it was snared and exteriorized via the femoral vein (Figure 4C & D). We advanced a 7 Fr. balloon wedge catheter through the venous side into the distal part of the fistula (Figure 4E). With the balloon inflated, we obtained much better images of the left coronary system, and could appreciate a distal branch (Figure 4F). Then we delivered the balloon wedge catheter into the left main coronary artery and introduced it into the nose of an 8 Fr. guiding catheter (kissing catheter technique) (Figure 4G). This allowed us to exchange a coronary wire for a 260-cm Glidewire® to form an AV loop (Figure 4H). We engaged a 6 Fr. AGA® delivery system, from the venous side into the mid LCx (Figure 4I & J). We chose

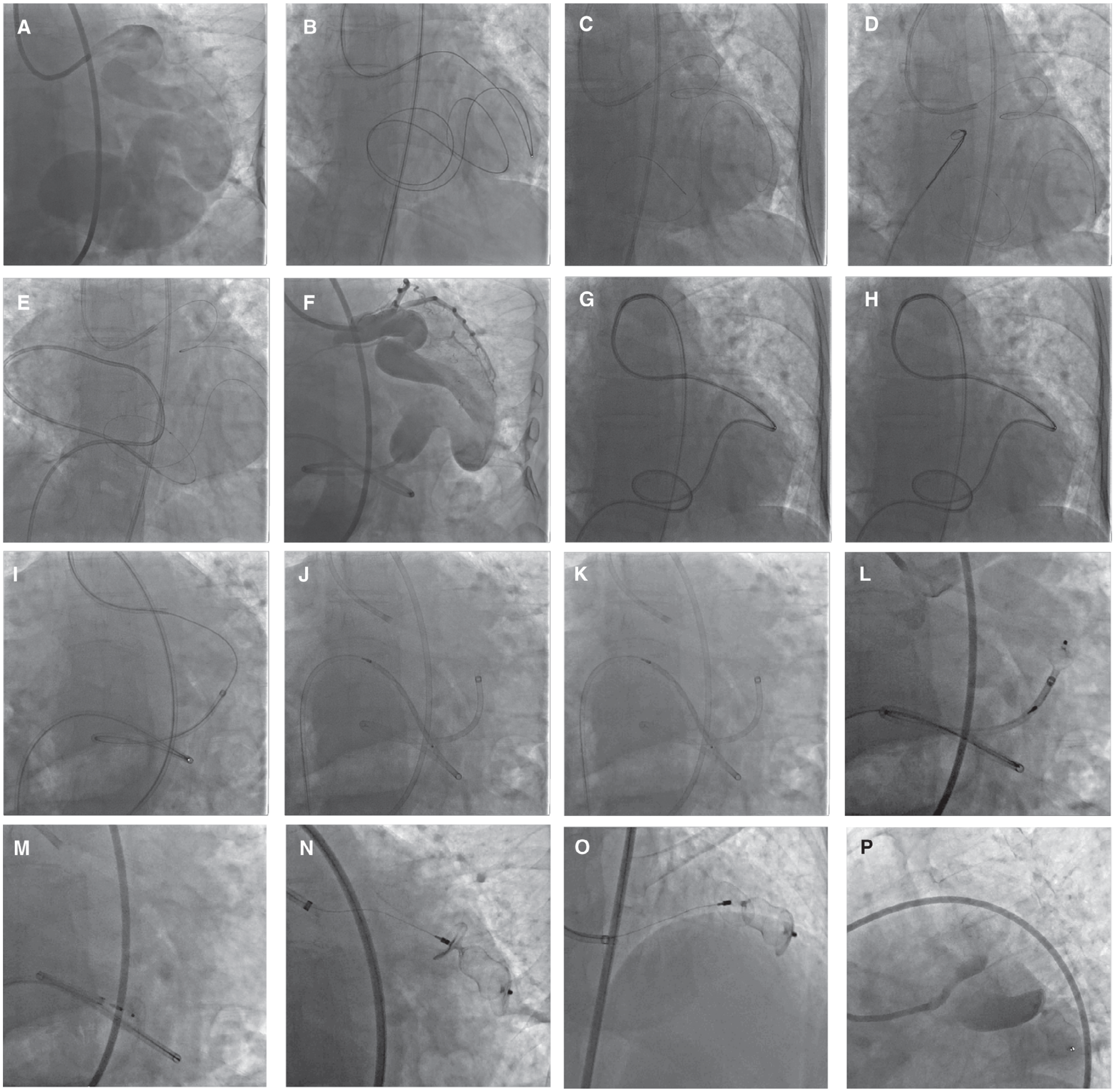

Figure 4. Arterio-venous. *Panel A*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.13. *Panel B*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.14. *Panel C*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.15. *Panel D*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.16. *Panel E*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.17. *Panel F*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.18. *Panel G*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.19. *Panel H*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.20. *Panel I*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.21. *Panel J*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.22. *Panel K*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.23. *Panel L*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.24. *Panel M*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.25. *Panel N*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.26. *Panel O*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.27. *Panel P*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.28.

to place a 12-mm Amplatzer Vascular Plug® in the middle portion of the fistula, as we calculated the distal fistula to be approximately 10 mm in diameter. The CS had multiple loops, resulting in poor torque control and an inability to precisely withdraw the catheter slowly. The device slipped back into the CS (Figure 4M). We then changed our strategy to a retrograde approach, due to the difficulty we encountered with the previous attempt. We once again made an AV loop, as demonstrated above and passed an 8 Fr. AGA® delivery system up to the mid LCx over the stiff exchange length Glidewire® to deliver an 18-mm Amplatzer Vascular Plug II® to successfully close the fistula (Figure 4N & P). As there was a branch originating beyond the site of occlusion, we left the device attached for about 15 minutes and released it only when patient remained asymptomatic and there were no changes on electrocardiogram.

This case highlights the complexities involved in closing large and tortuous CAFs, even through venous side. Often, 0.035-inch wires are used to make an AV loop to offer increased support for larger delivery sheaths. Waiting for a while before releasing the device, to assess for evidence of myocardial ischemia was important. The device could have been retrieved, if the patient had any chest pain or the ECG demonstrated ischemic changes.

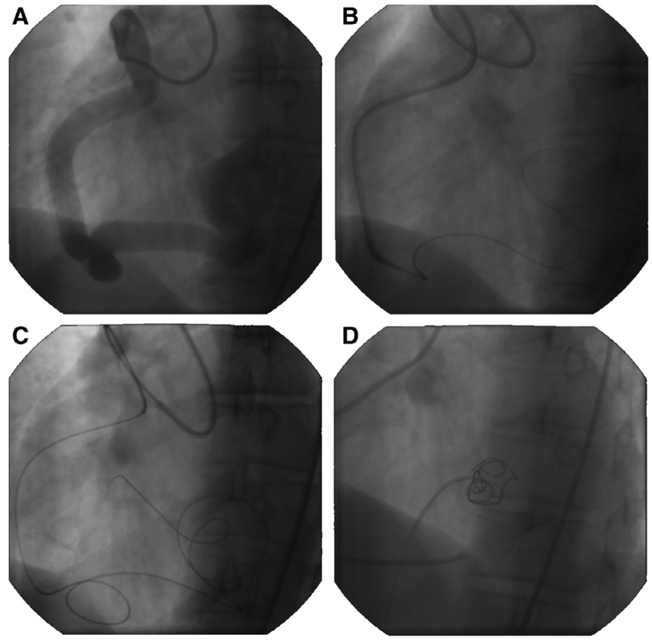

Figure 5. Retrograde approach – AV loop. *Panel A.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.29. *Panel B.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.30. *Panel C.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.31. *Panel D.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.32.

Retrograde Approach with an AV Loop

Here we demonstrate a large distal fistula from the RCA to the CS. Diagnostic angiography was performed using a 6 Fr. AL2® guide (Figure 5A). An initial attempt of using 5 Fr. balloon wedge catheter over a wire into the distal RCA failed (Figure 5B), so we used an 8 Fr. AL2® guide and 5 Fr. multipurpose catheter in a 'mother-child' format into the distal RCA. We used a 0.035-inch exchange length Glidewire® via the multipurpose catheter to exit the CS into the RA (Figure 5C). From there, it was snared and exteriorized through the femoral vein. We failed to advance any guide catheter through venous side into the CS, even using a kissing catheter technique over an AV loop. Eventually we advanced a 6 Fr. coronary guide catheter over the AV loop into the distal RCA from the retrograde approach, using the kissing catheter technique with a 4 Fr. SlipCath® in the inferior vena cava extending into the CS. Once we were satisfied with the guide catheter position in the distal RCA, we used a 150-cm Microferret® catheter (Obex, Auckland, New Zealand) to deliver platinum embolization coils to successfully occlude the flow. The catheter was kept stabilized in the distal RCA with help of an AV loop (Figure 5D).

Percutaneous Coronary Intervention Training

TCC of CAFs is a challenging coronary intervention that can result in coronary complications. Having percutaneous coronary interventional expertise not only offers additional skills to perform the procedure, but also is advantageous in managing complications. Anomalous coronary origins are not uncommon, and sometimes finding a catheter that offers adequate engagement and support is a challenge. The case below demonstrates the importance of a broad range of interventional cardiology skills training.

Case

A 55-year-old female presented with worsening exertional dyspnea. She was found to have two separate CAFs originating from proximal part of the RCA that had an anomalous origin and intra-arterial course. It was very difficult to obtain adequate catheter engagement, necessary for the procedure. We started with what we thought was going to be the best guide, an AL2®; however, this sat poorly on the aortic valve and kept prolapsing (Figure 6A). We tried a number of different guides; eventually, a JL® 3.0 guide engaged in the RCA ostium, although the support it offered remained poor (Figure 6B). The catheter prolapsed at the time of attempted coiling (Figure 6C). Using coils that can be retracted in such circumstances is very helpful. So, we placed a Prowater® coronary guide wire into the distal RCA, as an anchor wire to stabilize the catheter position (Figure 6D). This oriented the guide better and allowed us to use a Renegade® micro-catheter (Boston Scientific Inc., USA) over a Pilot® 50 wire (Abbott Vascular Inc., USA) to enter the distal fistula. We deployed four platinum

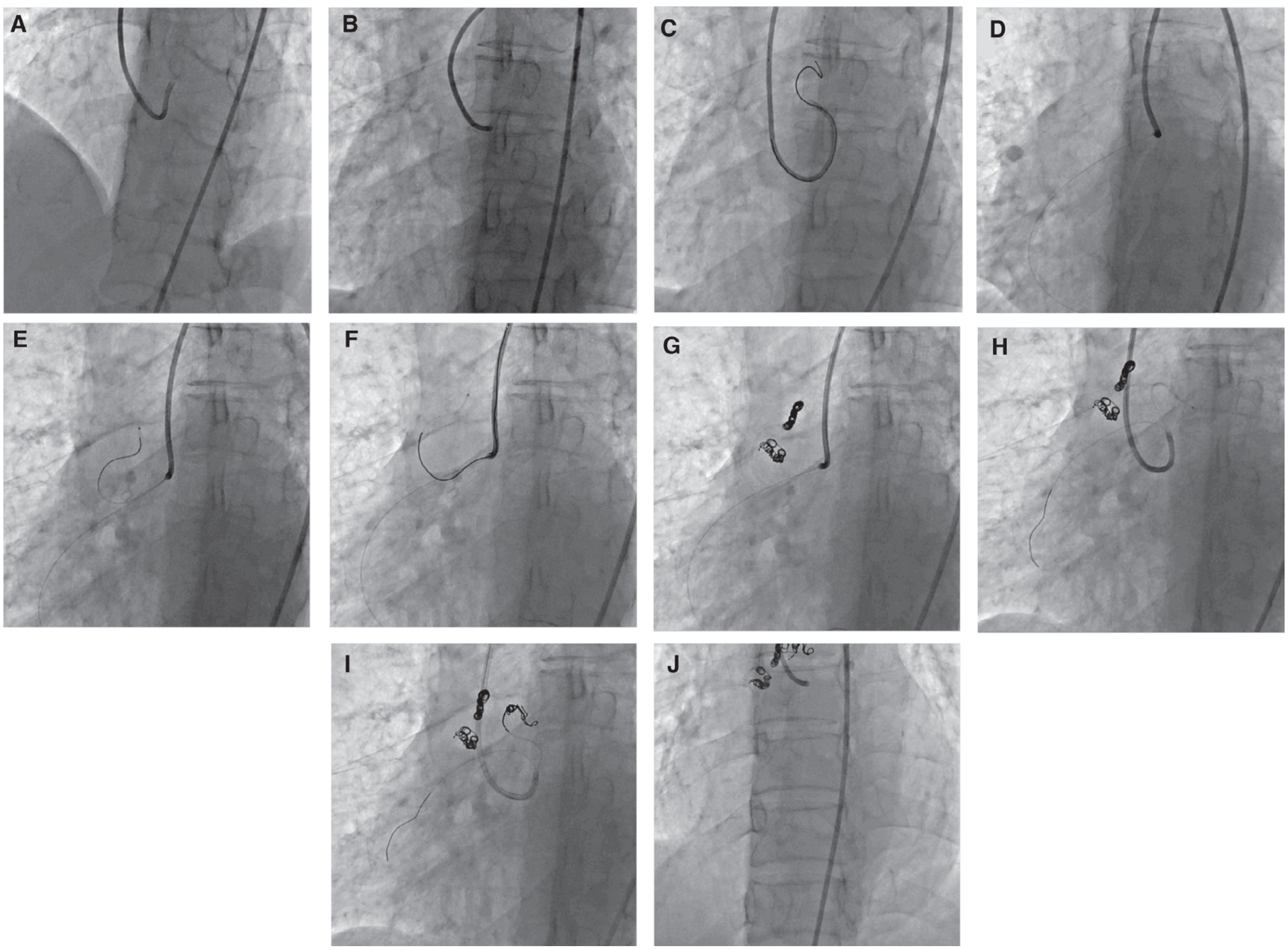

Figure 6. Important to have percutaneous coronary intervention skills. *Panel A.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.33. *Panel B.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.34. *Panel C.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.35. *Panel D.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.36 *Panel E.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.37. *Panel F.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.38. *Panel G.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.39. *Panel H.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.40. *Panel I.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.41. *Panel J.* View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.42.

interlock detachable IDC® coils measuring 3 mm × 10 cm to successfully close the CAF (Figure 6E & G). We then repositioned the guide allowing it to point upwards so we could wire the very proximal fistula. We were able to get the micro-catheter just proximal to the origin of the aneurysmal segment and deployed further IDC® coils, to successfully occlude all the fistulas (Figure 6H-J). The anchor wire helped us stabilize the guiding catheter position to achieve the final result.

Multiple Coronary Artery Fistulas

The majority of CAFs have a single feeding vessel but multiple feeding vessels are not rare (Figure 7A). Defining fistula branches and noting where they come off the coronary circulation is very important to understand before embarking on closure. As demonstrated in this case, it is important to systemically target as many feeding branches as possible to allow for vessel occlusion (Figure 7E & F).

Complications

TCC of CAF is the treatment of choice today, with success rates at par with surgically performed procedures, but with lower morbidity and mortality [1, 18]. Commonly encountered complications during TCC are as below,

Branch Occlusion

It is critical to adequately visualize distal branches which may be occluded or at risk of occlusion during the procedure. If a significant branch is at risk of occlusion, one should carefully consider the risk and benefits of the procedure.

ECG Changes and Arrhythmias

Careful monitoring of the 12 lead ECG during test occlusion and device deployment is critical to maintain safety. Our normal practice is to observe patients for the first 48 hours after fistula closure in a closely monitored environment, prior to considering for step-down. We do not anti-coagulate proximal fistulae, but routinely use heparin intra-operatively and warfarin after closure for large distal fistulae, where stasis may occur after successful occlusion.

Device Embolization

Coils and devices used to close CAF may dislodge and embolize at the time of deployment in a large fistula, due to high velocity flow. In a majority of reported cases coils could be retrieved, and no immediate complication were reported [19, 20].

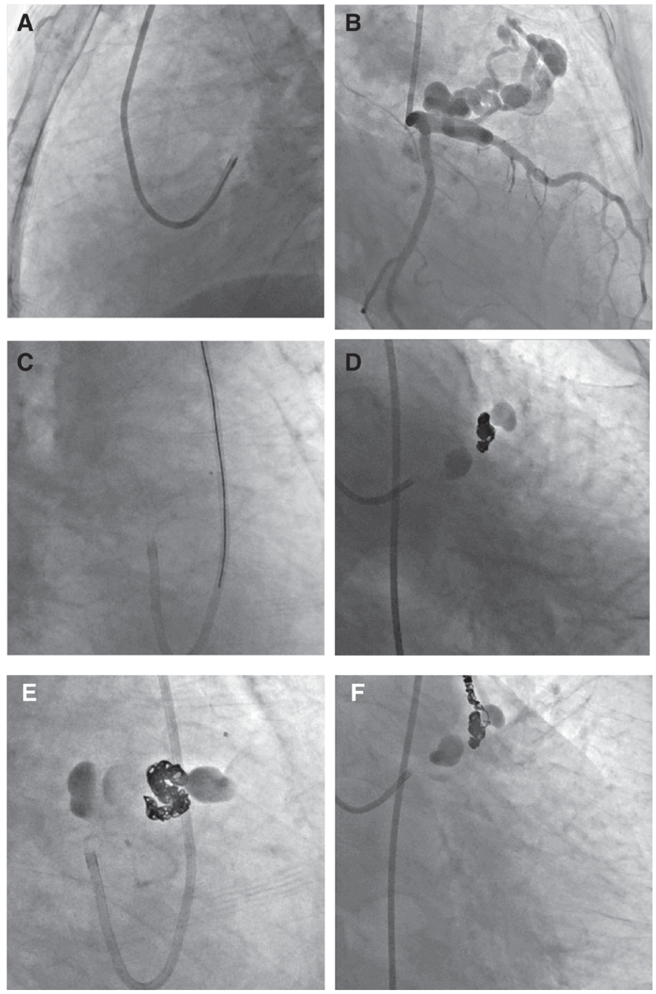

Figure 7. Multiple CAF. *Panel A*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.43. *Panel B*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.44. *Panel C*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.45. *Panel D*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.46. *Panel E*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.47. *Panel F*. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.013.14.vid.48.

Fistula Thrombosis

Complete closure of large CAFs results in a *cul-de-sac* within the vasculature with stagnant blood flow and poor native vessel runoff, creating an environment for thrombus formation. In a series of 16 patients, distal type, large-sized fistula and old age at the time of presentation were considered high risk factors leading to coronary thrombosis after CAF closure [21].

Thrombus Embolization

Thrombus formation proximal to the device is not uncommon. In some cases, thrombus can propagate proximally and embolize into adjacent coronary vasculature, resulting in ischemic complications. We believe this is more likely to happen, if coils or devices are deployed into the very proximal part of the CAF. It is important to deploy the implants far from branches of coronary arteries to prevent thrombus embolization. Our preferred strategy is to deliver device through an antegrade approach.

Myocardial Ischemia and Infarction

Myocardial damage at the time of a TCC is often multifactorial. Compromise of flow in branches of the coronaries beyond the point of occlusion is the most common cause. Thrombus formation proximal to the occlusion point can propagate or embolize into proximal branches. Device, catheter, and wire passage at the time of the procedure can damage coronary vasculature [20]. One should keep close watch for such potential complications, especially when tortuosity requires aggressive techniques. Sequential troponin measurement and transthoracic echocardiograms after CAF closure can be valuable to assess possible myocardial damage. There should be a very low threshold for performing repeat coronary angiography, if there are any concerns with myocardial ischemia.

Long-Term Follow Up and Unanswered Questions

Follow up after successful surgical or percutaneous closure of the CAF has been reported in a few case series with small numbers. In most series, patients have been followed only clinically and they have been found to be asymptomatic. Residual leaks, thrombosis of the CAF, myocardial ischemia/myocardial infarction, persistent coronary dilatation, and death have been reported after initially successful percutaneous CAF closure [18, 21-23].

Angiographic follow up at 1.5 years in a group of patients who have undergone TCC demonstrated 44% of the patients had recanalization of the fistulae and half underwent re-intervention to achieve complete TCC, suggesting angiographic follow-up may be necessary in this cohort [24]. There is very limited long-term angiographic and clinical follow up data in the literature. It is not clear whether CAF closure results in prognostic benefit to the majority of these patients.

Take Home Message

- Adults are more symptomatic than children with CAF.
- Adults are more prone to rupture and atherosclerotic disease.
- Large CAF and symptomatic patients should be considered for closure procedure.
- Small CAF and those with atypical symptoms should not be considered for closure.
- CAF closure is not a routinely performed procedure.
- Surgical consultation should always be obtained.
- It is rare to find a significant Qp:Qs with CAF.
- Fully anticoagulate patients to avoid thrombotic complications.
- Plan a ‘tailor-made’ strategy for each individual case.
- Make sure to have various devices on the shelf.
- Necessary to have angioplasty skills readily available (yourself or a colleague).
- Be gentle with devices to avoid coronary complications.
- Careful evaluation for any potential complication before closing the case.
- Low threshold for repeat angiography if there is recurrence of symptoms, ECG changes, or new onset regional wall motion abnormality.

- Limited evidence is available on long-term outcome and prognostic benefit of the closure procedure.

Conflict of Interest

The authors have no conflict of interest relevant to this publication.

Comment on this Article or Ask a Question

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Pitfalls of Echocardiography Diagnosis of Anomalous Origin of Left Coronary Artery from the Main Pulmonary Artery

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Abstract

A 14-year-old female was referred by her pediatrician for evaluation of chest pain. Most of her chest pain was experienced during school gym class, limiting her participation. No history of syncope was found. She had never been to the emergency room with chest pain. She has always been in good health. Physical examination was normal. A 2D echocardiogram was misleading; however, a color flow Doppler echocardiogram confirmed the diagnosis of anomalous origin of the coronary artery from the main pulmonary artery (ALCAPA). She underwent successful surgical correction with excellent results.

Key Words

Anomalous origin of left coronary artery • Chest pain • 2D-color Doppler echocardiogram

Introduction

Chest pain in children is a very common presentation in a busy pediatric cardiology practice as well as in the emergency room. The importance of taking a thorough history with respect to exertional chest pain in children cannot be overemphasized. Our purpose is to present a 14-year-old healthy female who presented in our clinic with a history of recurrent chest pain after physical activities. A 2D echocardiogram was misleading, as if the left coronary artery (LCA) arose from the left coronary sinus; however, color Doppler flow reversal within the LCA confirmed the diagnosis of anomalous origin of the coronary artery from the main pulmonary artery (ALCAPA). Our purpose is to emphasize the importance of color Doppler flow patterns within the coronary arteries to confirm their origins. A 2D echocardiogram alone could be misleading.

The incidence of ALCAPA is 1 in 300,000 live births [1]. Infants usually present with severe congestive heart failure at around 2 months of age when the pulmonary artery pressure decreases. Abdominal colic with feeding that mimics angina pectoris from poor myocardial perfusion can be an earlier presentation.

Our experience strongly emphasizes that only imaging the origin of the LCA from the left coronary sinus by a 2D echocardiogram alone is not enough to exclude this diagnosis [2]. Our patient exemplifies the importance of color flow Doppler imaging to confirm the diagnosis of ALCAPA.

Case Report

A 14-year-old Hispanic female complained of exertional, nonradiating recurrent chest pain over the left precordium. She described the chest pain as "pinching" in nature and used to get relief with rest. Chest pain had limited her school gym class physical activities. She denied any associated symptoms such as pre-syncope, syncope, or palpitations. She had never been to the emergency room for her chest pain.

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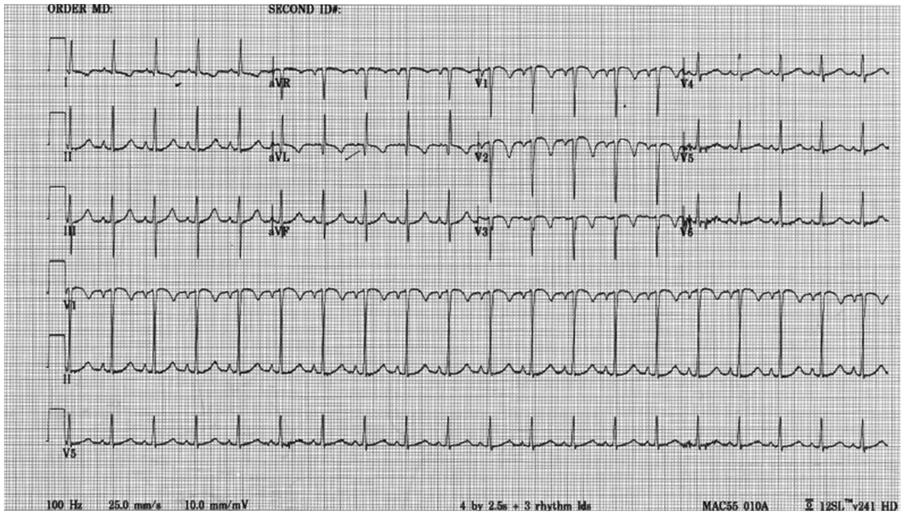

Figure 1. Twelve-lead electrocardiogram showing inverted T in lead I and avL.

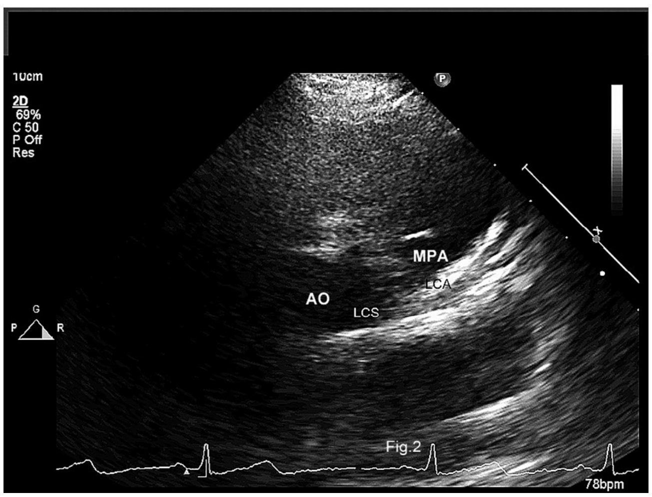

Figure 2. A 2D echocardiogram. The LCA appears to arise from the LCS. AO = aorta; LCA = left coronary artery; LCS = left coronary sinus; MPA = main pulmonary artery.

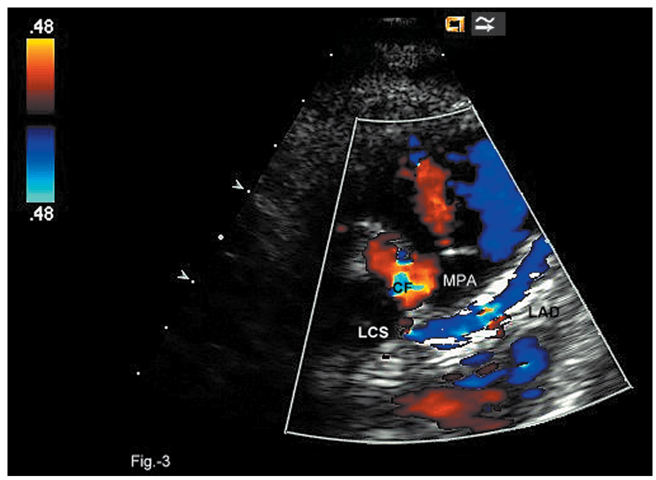

Figure 3. Color flow Doppler mapping showing reversal of color flow within the LCA (blue color). CF =coronary flow; LCA = left coronary artery; LCS = left coronary sinus; MPA = main pulmonary artery.

Her physical examination was unremarkable. Vital signs were heart rate 71/min, respiratory rate 18/min, and blood pressure 110/56 mm Hg. Peripheral pulses were normal and equal in the upper and lower extremities. Cardiac examination revealed normal first and second heart sounds. There was no audible murmur, pericardial rub, or click.

An electrocardiogram (Figure 1) showed normal sinus rhythm and no evidence of chamber hypertrophy. Inverted T in lead I and AVL suggested LV strain. Significant q in aVL was also seen. No ST segment changes were seen.

2D imaging was misleading, as if the LCA arose from the left coronary sinus (Figure 2). However, color Doppler showed reversal of color flow within the LCA (Figure 3) from retrograde filling from the dilated right coronary artery, measuring 4.5 mm with a z-score of +3.14 (Figure 4) by the collateral vessels, suggesting a diagnosis of ALCAPA. This prompted us to explore further with angiography, confirming our diagnosis of ALCAPA (Figures 5 and 6). Subsequently, she underwent corrective surgery with LCA implantation into the aorta with excellent results.

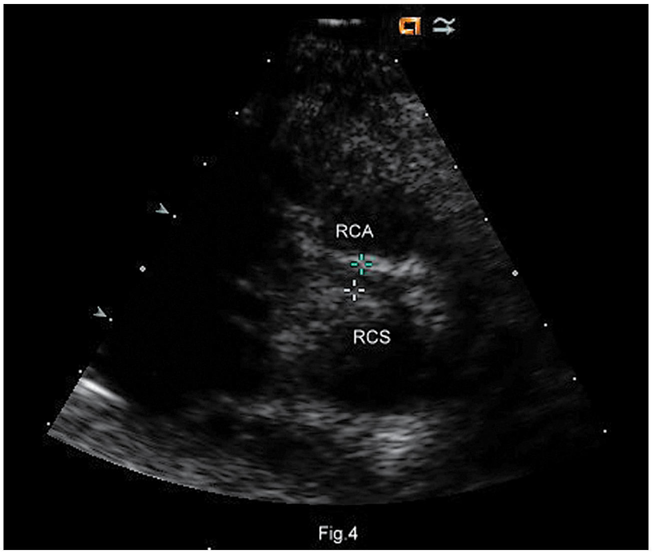

Figure 4. Dilated RCA is arising normally from the RCS. RCA = right coronary artery; RCS = right coronary sinus.

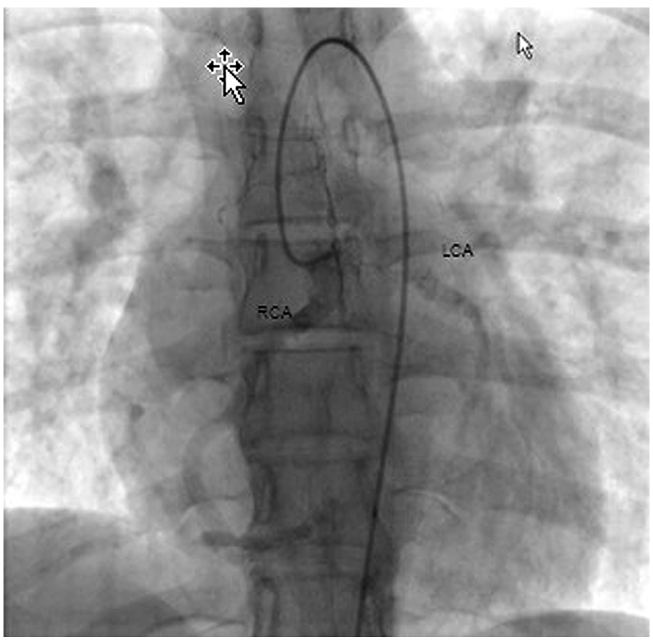

Figure 5. RCA angiogram in the P-A view, showing retrograde filling of the LCA by collaterals . LCA = left coronary artery; RCA = right coronary artery.

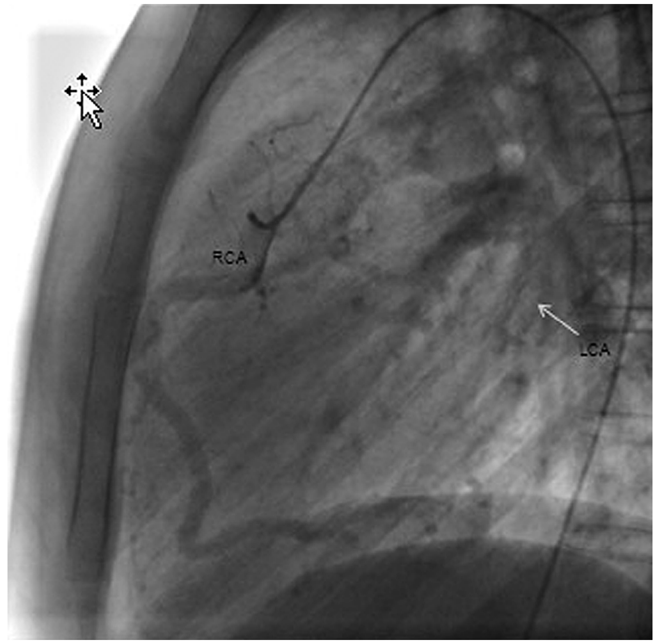

Figure 6. RCA angiogram in the lateral view, showing retrograde filling of the LCA by collaterals (arrow). LCA = left coronary artery; RCA = right coronary artery.

Conflict of Interest

The authors have no conflicts of interest relevant to this publication.

Comment on this Article or Ask a Question

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State-of-the-Art Periprocedural 3D Transesophageal Echocardiography during Transcatheter Mitral Valve-in-Valve Implantation

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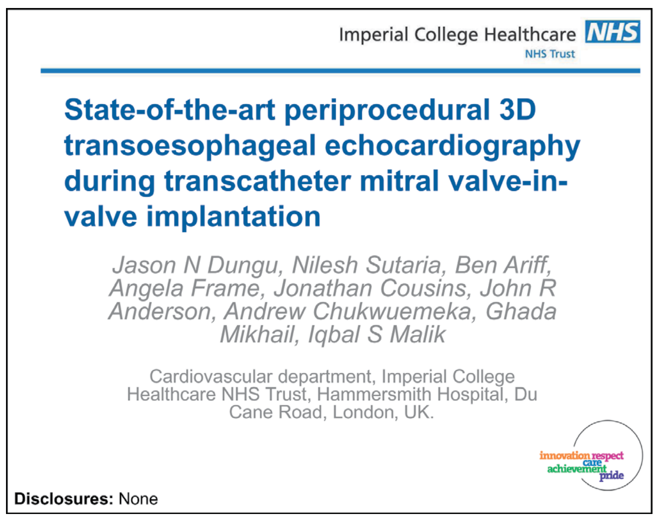
Abstract

An 88 year old lady, with a previous 25 mm Carpentier-Edwards 6900 Perimount pericardial mitral bioprosthesis in 2007 for severe mitral regurgitation, presented in September 2013 with shortness of breath on mild exertion with New York Heart Association class 3 heart failure. Transthoracic and transoesophageal echocardiography demonstrated good biventricular systolic function but significant transvalvular mitral prosthesis regurgitation and severe restriction of the leaflets. She was reviewed by the cardiothoracic surgeons and turned down for re-do mitral valve surgery due to frailty. The mitral multi-disciplinary team recommended transcatheter mitral valve-in-valve implantation.

A temporary pacing wire was inserted via the right internal jugular vein. The procedure was performed via transapical access with a 6F sheath. A standard J-wire crossed the mitral valve easily with the aid of a pigtail catheter. The Transcatheter Aortic-Valve Implantation delivery sheath was positioned across the mitral valve prosthesis, guided by 2D and real time 3D transoesophageal echo (TOE) and fluoroscopy. A 26 mm Edwards XT Ascendra Transcatheter Valve (Edwards Lifesciences, Irvine, California) was deployed successfully under rapid pacing within the bioprosthetic ring. A good position was seen on fluoroscopy and 2D and 3D TOE showed only minimal paravalvular leak. A significant increase in mitral valve area and reduction in transmitral gradient was observed. The patient was discharged on day 6 and remains well at 6 months review.

Valve-in-valve implantation, a new technology only possible with state-of-the-art imaging, is a viable treatment option for degenerative tissue bioprosthesis disease in high risk surgical patients.



Slideshow. Click on the presentation above to view the presentation online. You may download the file at http://dx.doi.org/10.12945/j.jshd.2016.002.14.ppt.01 (5.6MB).

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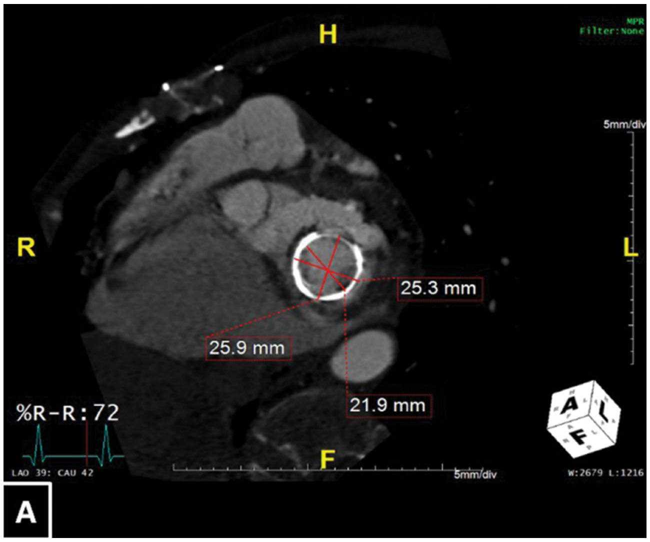


Figure 1. Sizing of the prosthetic annulus preprocedure by computed tomography, confirming maximum diameter of 25.9 mm.

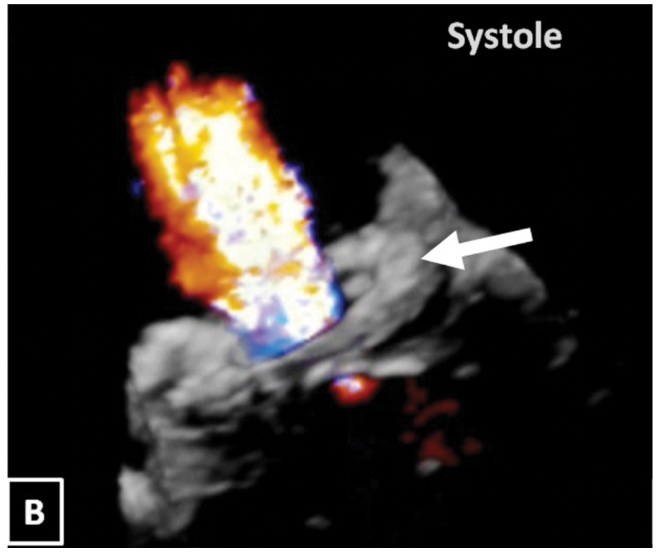
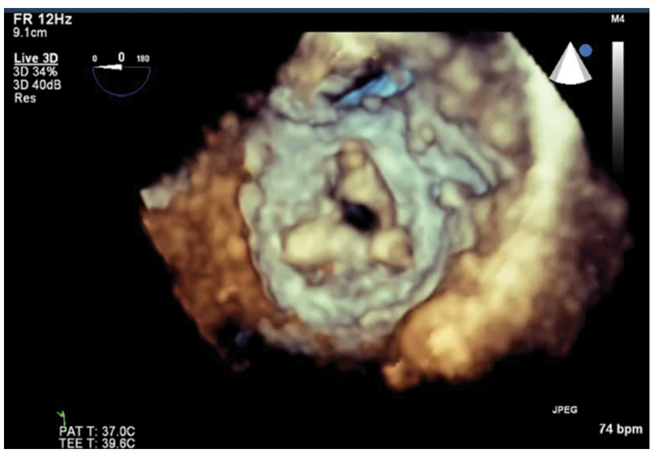
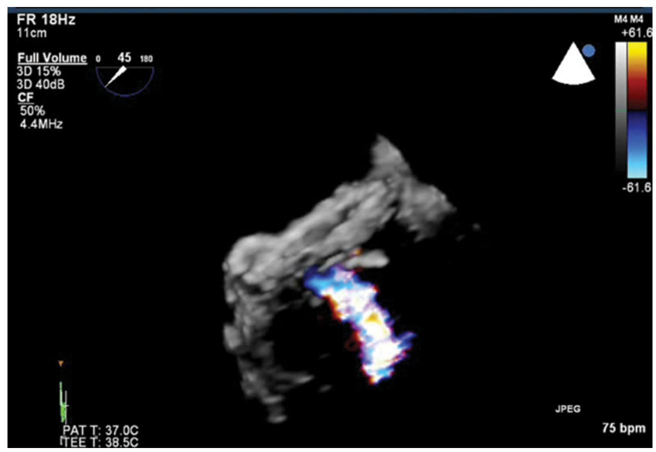


Figure 2. A 3D transoesophageal echo full volume colour in systole, demonstrating severe transvalvular mitral regurgitation.



Video 1. Real-time 3D zoom (left ventricular view) preintervention, demonstrating severely restricted bioprosthetic leaflet opening. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.002.14.vid.01.



Video 2. Full volume 3D color Doppler preintervention, demonstrating severe transvalvular regurgitation and turbulent diastolic flow due to severe mitral bioprosthesis stenosis. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.002.14.vid.02.

Key Words

Valve-in-valve • Mitral bioprosthesis • TAVI • Percutaneous intervention • 3D transoesophageal echocardiography • General

Transapical transcatheter mitral valve-in-valve implantation was first reported in 2009 [1] and small case series have been reported [2, 3]. We present a case of transcatheter aortic valve implantation (TAVI) in the mitral position, highlighting the role of 3-dimensional echocardiography during this complex intervention in the cardiac catheter laboratory.

An 88-year-old lady, with a previous 25-mm Carpentier-Edwards 6900 Perimount pericardial mitral bioprosthesis in 2007 for severe mitral regurgitation, presented in September 2013 with shortness of breath on moderate exertion with New York Heart Association (NYHA) class 3 heart failure. Transthoracic

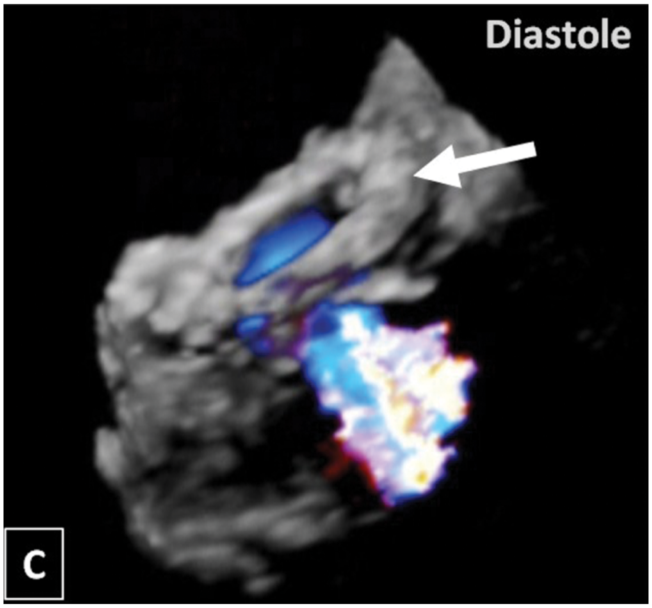


Figure 3. A 3D transoesophageal echo full volume colour in diastole, demonstrating flow convergence and turbulence secondary to mitral prosthesis stenosis. Solid arrow: Mitral valve prosthesis.

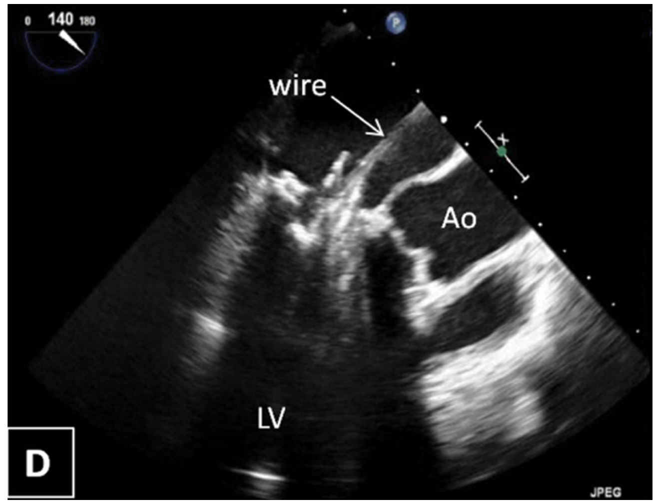
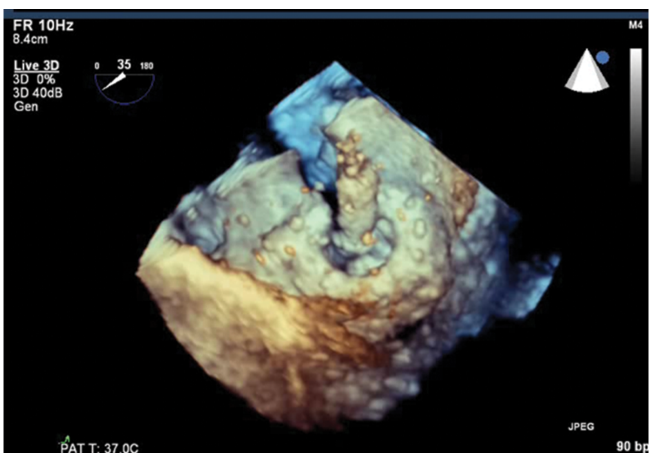
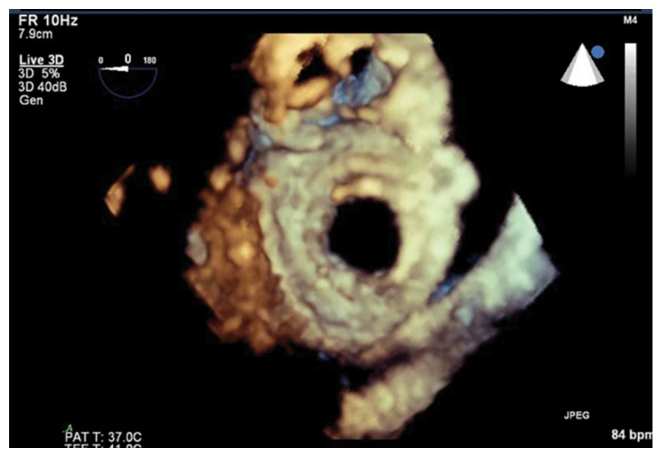


Figure 4. A 2D transoesophageal echo (LVOT - view 140 degrees) with the wire across the mitral valve into the left atrium. LV = Left ventricle; LVOT = left ventricular outlow tract; Ao = Aorta.



Video 3. Live 3D transoesophageal echo-guided positioning of transcatheter aortic valve implantation in the mitral position. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.002.14.vid.03.



Video 4. Live 3D left ventricular view post transcatheter aortic valve implantation in the mitral position. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.002.14.vid.04.

and transesophageal echocardiography demonstrated good biventricular systolic function but significant transvalvular mitral prosthesis regurgitation and severe restriction of the leaflets. Left and right heart catheterization demonstrated unobstructed epicardial coronary arteries and severely elevated peak pulmonary artery systolic pressure (PASP) of 80 mm Hg. She was reviewed by the cardiothoracic surgeons and turned down for repeat mitral valve surgery due to frailty. The mitral multidisciplinary team decision for transcatheter mitral valve-in-valve implantation was subsequently discussed with the patient who accepted a 10% risk of death or major adverse cardiac event. Sizing of the prosthetic annulus and leaflet area was performed pre-procedure using computed tomography (CT) (Figure 1) and a plan for 26-mm device implantation was made.

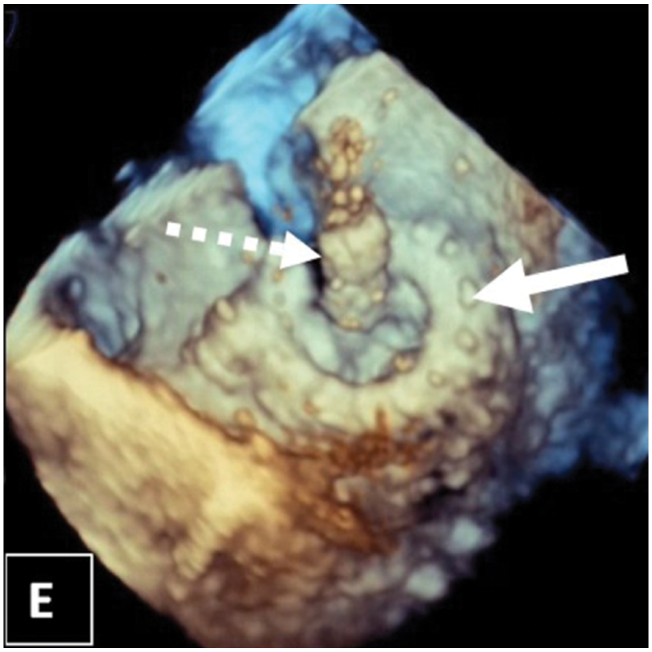


Figure 5. Real time 3D zoom transoesophageal echo during position of transcatheter aortic valve implantation in the mitral position. Solid arrow = Mitral valve prosthesis; dashed arrow = 26-mm Edwards TAVI.

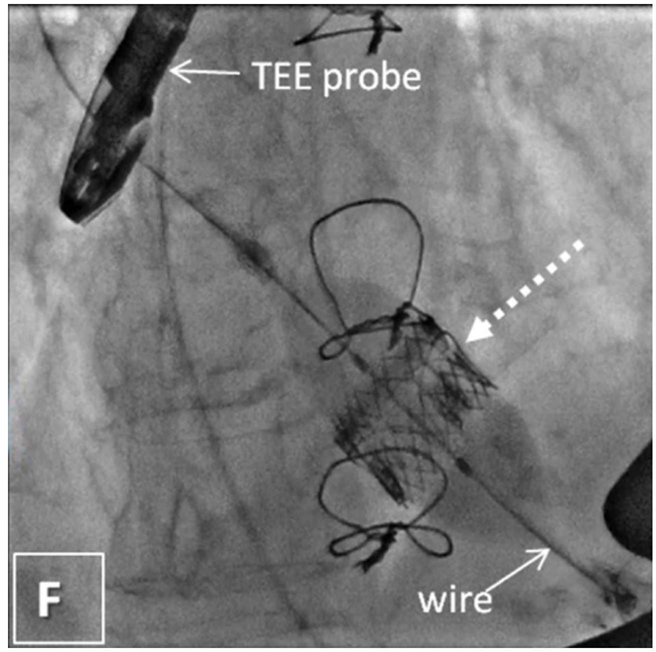
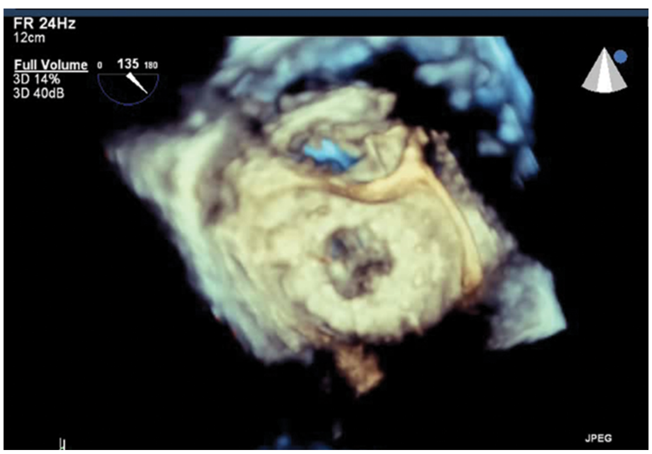
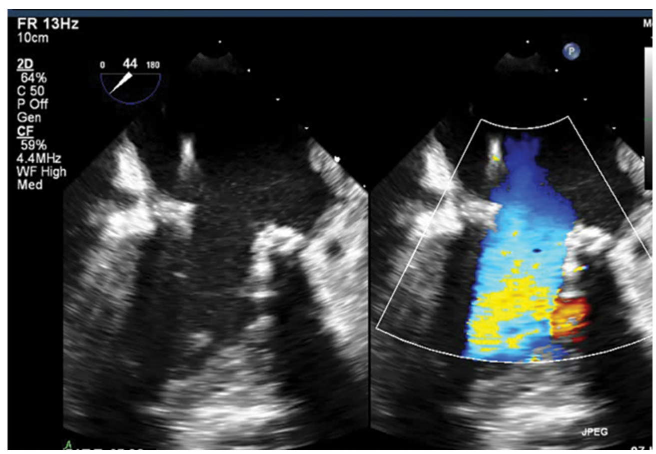


Figure 6. Deployment of transcatheter aortic valve implantation (TAVI) in mitral position (fluoroscopy). dashed arrow = 26-mm Edwards TAVI.



Video 5. 3D full volume left atrial view postintervention. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.002.14.vid.05.



Video 6. 2D transoesophageal echo colour compare postintervention, demonstrating good transcatheter aortic valve implantation in mitral position function. View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.002.14.vid.06.

Procedural Details

A 3D zoom transoesophageal echo (TOE) (left ventricular view) pre-intervention demonstrated severely restricted bioprosthetic leaflet opening (Video 1). Full volume 3D TOE with colour Doppler revealed severe transvalvular mitral prosthesis regurgitation with a broad systolic jet into the left atrial (LA) and flow turbulence on the ventricular side in diastole indicative of severe stenosis of the prosthesis (Figure 2, Figure 3, and Video 2). A temporary pacing wire was inserted via the right internal jugular vein. The procedure was

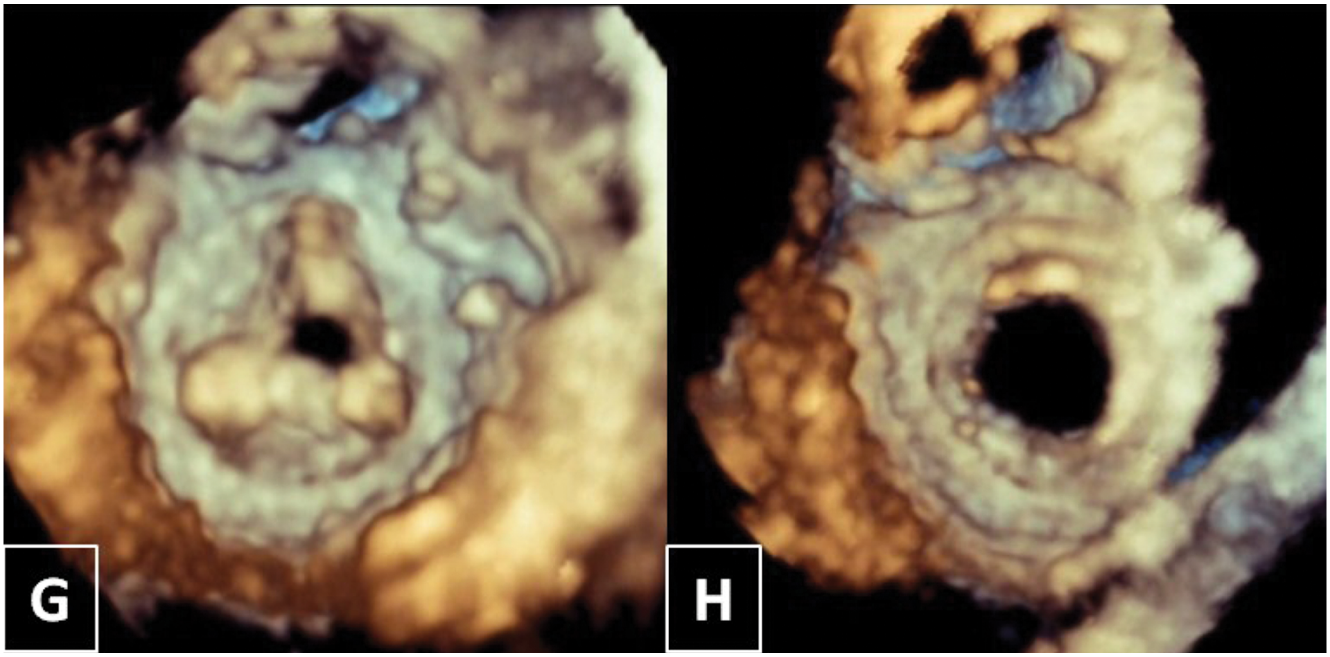


Figure 7. Real time 3D zoom (left ventricular view) in diastole, demonstrating the increase in mitral valve area postintervention.

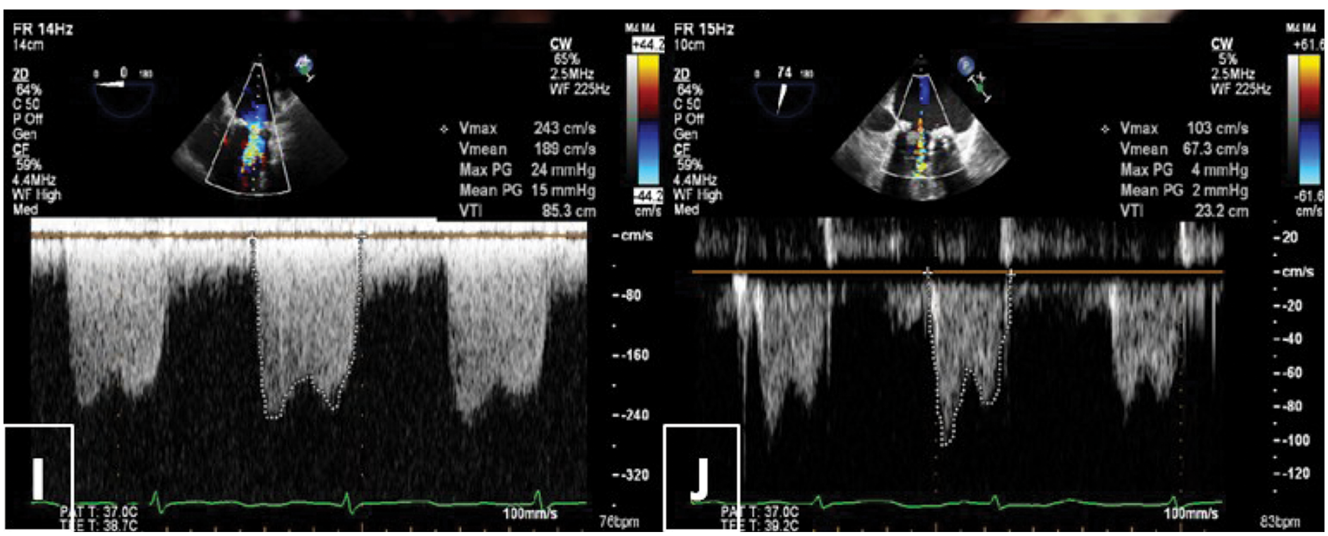
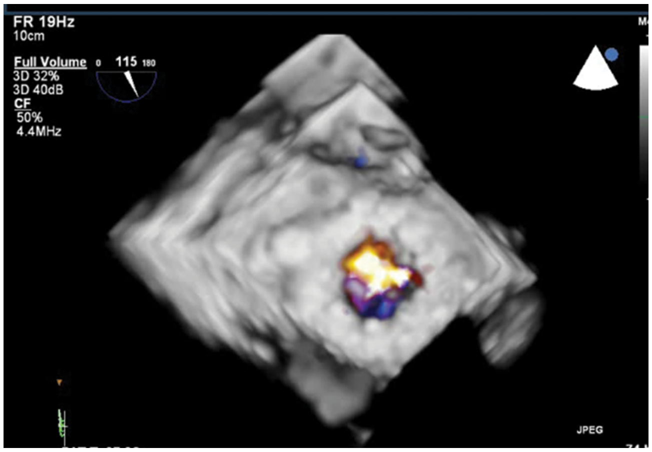


Figure 8. Continuous wave Doppler, demonstrating the reduction in mean mitral valve gradient (preintervention 15 mm Hg; postintervention 2 mm Hg).

performed via transapical access with a 6F sheath. A standard J-wire crossed the mitral valve easily with the aid of a pigtail catheter. A 26-mm Edwards XT Ascendra Transcatheter Valve (Edwards Lifesciences, Irvine, California, USA) delivery sheath was positioned across the mitral valve prosthesis, guided by 2D and real time 3D TOE and fluoroscopy (Figure 4, Figure 5, and Video 3). The transcatheter valve was deployed successfully under rapid pacing within the bioprosthetic ring (Figure 6). A good position was seen on fluoroscopy and 2D and 3D TOE showed only minimal paravalvular leak (Videos 4-7). A significant increase in mitral valve area and reduction in transmitral gradient was observed (Figures 7-8). The patient remained hemodynamically stable throughout the procedure. The temporary wire was removed and the patient was transferred to the cardiothoracic intensive care unit and discharged on day 6 on aspirin and clopido-



Video 7. Full volume 3D color Doppler postintervention (left atrial view). View supplementary video at http://dx.doi.org/10.12945/j.jshd.2016.002.14.vid.07.

grel. She remained well at 6 month review with a well seated valve on transthoracic echocardiography and a reduction in estimated pulmonary artery systolic pressure to 46mmHg.

Valve-in-valve implantation, a new technology only possible with state-of-the-art imaging, is a viable treatment option for degenerative tissue bioprosthesis disease in high risk surgical patients. CT is helpful for accurate sizing of the Edwards transcatheter stented valve and detailed periprocedural examination by TOE is needed to define anatomy, guide the procedure and provides immediate assessment of potential complications.

Conflict of Interest

The authors have no conflicts of interest relevant to this publication.

Comment on this Article or Ask a Question

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