

Transcatheter closure of complex iatrogenic ventricular septal defect: a case report

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Background

Iatrogenic membranous ventricular septal defects (VSDs) are rare complications of cardiothoracic surgery, such as septal myectomy for hypertrophic obstructive cardiomyopathy (HOCM). Transcatheter closure is considered an appealing alternative to surgery, given the increased mortality associated with repeated surgical procedures, but reports are extremely limited.

Case summary

We herein report the case of a 63-year-old woman with HOCM who underwent successful percutaneous closure of an iatrogenic VSD after septal myectomy. Two percutaneous techniques are discussed, namely the ‘muscular anchoring’ and the ‘buddy wire delivery’, aimed at increasing support and providing stability to the system during percutaneous intervention.

Discussion

Transcatheter closure represents an attractive minimally invasive approach for the management of symptomatic iatrogenic VSDs. The new techniques described could help operators to cross tortuous and tunnelled defects and to deploy closure devices in case of complex VSD anatomy.

Keywords

Hypertrophic obstructive cardiomyopathy • Ventricular septal defects • Percutaneous intervention • Case report

Learning points

- Septal reduction therapy decisions for symptomatic hypertrophic obstructive cardiomyopathy (HOCM) patients should be jointly made by a multidisciplinary team including cardiologists, interventional cardiologists, and expert cardiac surgeons.
- Complex iatrogenic ventricular septal defects are rare complications after septal reduction therapies in HOCM patients and can be challenging to treat. Percutaneous procedures could be an attractive less-invasive way to manage these complications in high-risk patients.
- Percutaneous closure devices are not intended for iatrogenic ventricular defects treatment, thus implantation can be cumbersome due to unfavourable anatomical characteristics.

Introduction

Medical treatment is the first-line therapy for symptomatic patients with hypertrophic cardiomyopathy (HCM) and left ventricular outflow tract (LVOT) obstruction. However, when medical treatment

fails, surgical myectomy or percutaneous alcohol septal ablation are regarded as standard therapies to eliminate subaortic obstruction.¹

Iatrogenic ventricular septal defects (VSDs) are rare complications after surgical treatment of hypertrophic obstructive cardiomyopathy (HOCM).² Due to the increased mortality associated with repeat surgery and the technological and operational improvement in

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transcatheter procedures, percutaneous closure of congenital, post-myocardial infarction, and iatrogenic VSDs has become the therapy of choice. However, there is little data concerning this approach for VSDs management after septal myectomy in HOCM.^{3,4} Therefore, we hereby report a case of successful percutaneous closure of an iatrogenic VSD following surgical therapy for HOCM.

Timeline

Day 0	Patient with history of hypertrophic obstructive cardiomyopathy referred to our institution due to symptoms' progression
Day 0	Physical examination, 12 lead electrocardiogram and transthoracic echocardiography were performed revealing signs and symptoms of heart failure due to severe left ventricular outflow tract obstruction
Day 3	Ventricular septal myectomy was performed
Day 3	Transoesophageal echocardiography revealed two ventricular septal defects (VSDs)
Day 7	Percutaneous VSD closure was performed
Day 7	Post-procedure transoesophageal echocardiography showed non-significant residual shunt

Case presentation

A 63-year-old woman, with a history of hypertension, Hashimoto's thyroiditis, and HOCM symptomatic for dyspnoea [New York Class Association (NYHA) III], was referred to our Institution for septal myectomy. The diagnosis of HOCM (Type III according to Maron's classification) was made 4 years before admission; the patient was only mildly symptomatic for dyspnoea (NYHA Class I-II) at that time, and the transthoracic echocardiogram (TTE) showed an interventricular septal wall thickness of 18 mm (normal range 6–10 mm) and an LVOT gradient of 70 mmHg. Since then, maximum tolerated medical therapy with beta-blocker (atenolol 100 mg q.d.), disopyramide (250 mg b.i.d.), and spironolactone was introduced, and regular outpatient follow-up was carried out. On admission, physical examination revealed an ejection systolic murmur on the left sternal edge, radiating to the apex, and increased by Valsalva manoeuvre. Other physical findings were pulmonary rales in the lower lung fields, mild distension of the jugular veins and lower extremity oedema. A 12-lead electrocardiogram showed sinus rhythm, atrial dilation, and giant T-wave inversion in the precordial leads (*Figure 1*). Basal TTE showed normal left ventricular (LV) function and confirmed the persistence of severe LVOT obstruction (peak outflow tract gradient 85 mmHg at rest). The presence of an LVOT obstruction with increased pressure gradient was associated with a systolic anterior movement of the mitral valve anterior leaflet, resulting in severe mitral regurgitation. The left atrium was only mildly dilated [left atrial (LA) indexed-volume 35 mL/m² (normal range 16–34 mL/m²), LA diameter 53 mm (normal range 27–38 mm)] (*Supplementary material online, Video S1–S3*). Left cardiac

catheterization and coronary artery angiography showed no significant coronary artery stenoses and a peak-to-peak LVOT gradient of 100 mmHg, with typical intensified gradient after premature ventricular contraction (Brockenbrough-Braunwald-Morrow sign), confirming LVOT obstruction.

As the patient was still in NYHA Class III with resting LVOT gradient of >50 mmHg despite maximum tolerated medical therapy, septal myectomy was planned by our Heart Team and performed at our high-volume institution, according to the latest guidelines.¹ The HCM 5-year risk score for sudden cardiac death (HCM-SCD score) of 2.94% did not mandate implantable cardioverter-defibrillator implantation.

Septal myectomy was extended from the nadir of the right aortic cusp to the commissure of the right and left coronary cusps, and carried down to the anterolateral papillary muscle level. Mitral valve intervention was not performed. Early after procedure, a VSD was evident and intraoperative repair with a 3 × 3 cm bovine pericardium patch was attempted. A residual small peri-apical VSD was observed after surgery. The following transoesophageal echocardiography revealed two VSDs: the widest (18 × 4 mm, area 6 mm²) was located at the distal edge of the myectomy, presented irregular rims and caused biventricular shunt, the smallest (max diameter 2 mm) was located in the membranous septum and did not cause significant shunt (*Figure 2* and *Supplementary material online, Video S4*).

Due to worsening symptoms and haemodynamic instability, a new interventional approach was required. Given the high risk of a re-do surgery, the Heart Team decision was to proceed with percutaneous closure of the VSD. A 25-mm AMPLATZER® Multi-Fenestrated Septal Occluder—'Cribiform' (St. Jude Medical-Abbott, Minneapolis, MN, USA) was chosen to close the wider defect (see *Supplementary material online, Video S5*) through a standard femoral antegrade approach. The defect was crossed from the LV side using a right coronary guiding catheter and a 0.035" hydrophilic wire, but the wire got entangled in the LV apical trabeculae. After multiple attempts with different catheters in order to increase support, we opted for a JR4 guiding catheter with a coronary 0.014" guidewire, which was advanced through the VSD. A 4.0 × 8 mm non-compliant balloon was then inflated into the narrow part of the defect to obtain a 'muscular anchoring', in order to push forward the catheter and allow support for advancing a 0.018" exchange length guidewire in the right ventricle (*Figure 3A* and *Supplementary material online, Video S6*). The wire was then snared in the pulmonary artery and externalized through the femoral venous sheath, creating an arteriovenous loop (*Supplementary material online, Video S7*). The delivery sheath was advanced over the wire from the femoral vein and pushed across the defect into the left ventricle, simultaneously pulling the arterial end of the wire to avoid bending (*Supplementary material online, Video S8*). However, since the tunnelled and tortuous nature of the VSD, the wire was left in place as a safety 'buddy' wire, in order to improve support and avoid the accidental withdrawal of the system. The delivery cable was introduced inside the delivery sheath, next to the wire, and advanced through the VSD. The device was then positioned and deployed, while holding the wire in tension during the whole procedure (*Figure 3B*). Finally, after guidewire removal, the device was released under echocardiographic guidance (*Supplementary material online, Video S9*). Echocardiography showed a good result with minimal

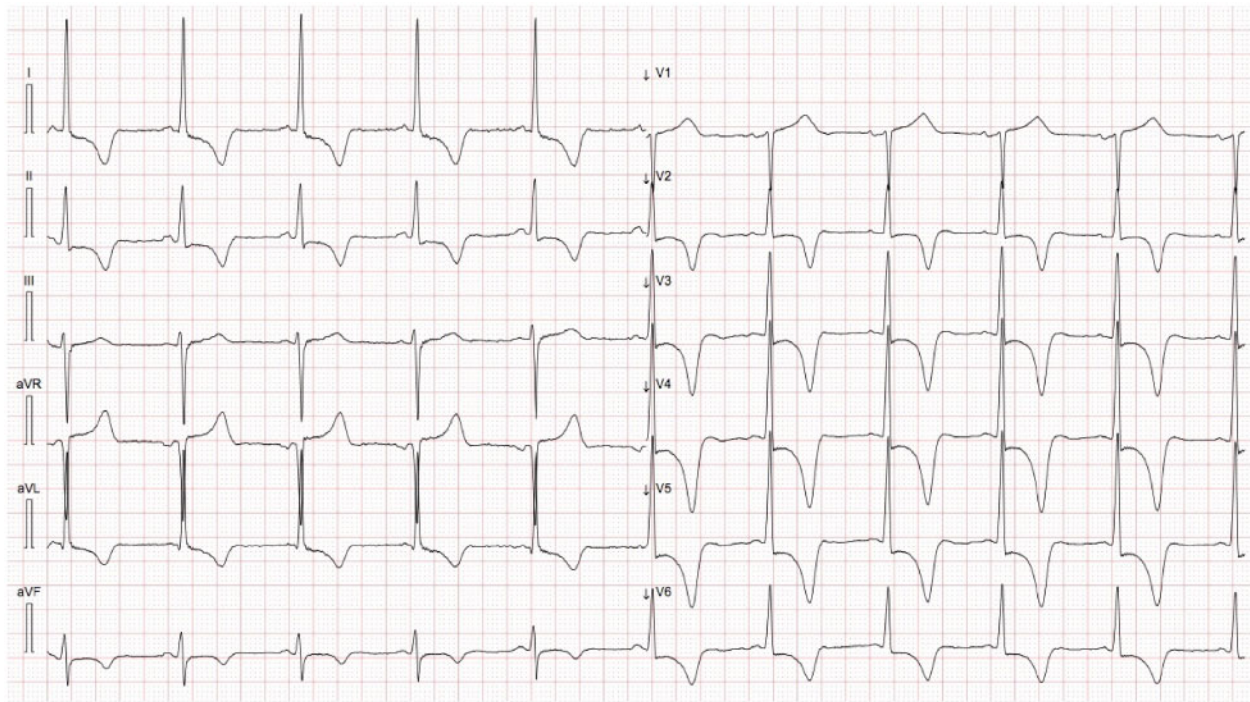


Figure 1 Baseline electrocardiogram on admission showing sinus rhythm, atrial dilation and giant T-wave inversion in the precordial leads.

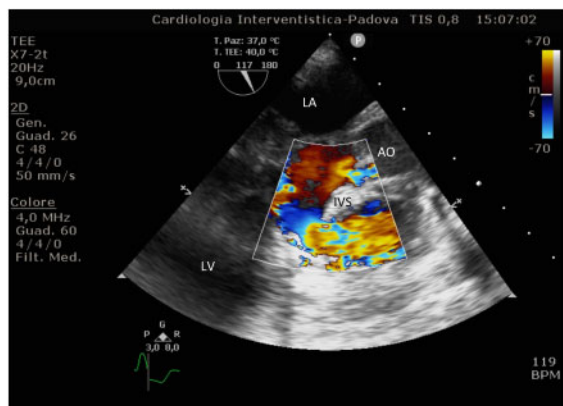


Figure 2 Pre-procedural transoesophageal echocardiography showing a wide ventricular septal defect located at the distal edge of the myectomy. AO, aorta; IVS, interventricular septum; LA, left atrium; LV, left ventricle.

residual shunt (Figure 4 and Supplementary material online, Video S10). At 30-day follow-up, echocardiographic findings were similar and the patient was only mildly symptomatic for exertional dyspnoea (NYHA Class I-II).

Discussion

When performed by experienced surgeons in high-volume centres, isolated septal myectomy has <1% mortality and low rates of major

complications, including heart block, aortic regurgitation, and VSD.² The American College of Cardiology/American Heart Association (ACC/AHA) Guidelines for the Diagnosis and Treatment of Patients with HCM have therefore recommended that septal reduction therapy should be performed only by experienced operators in the context of a comprehensive HCM clinical programme.⁵ In the unfortunate event of a complication such as a VSD, additional cardiac surgery to repair the defect can cause substantial morbidity and death, and transcatheter closure could be an attractive less-invasive option in high-risk patients. However, transcatheter closure devices are not specifically intended for implantation in such iatrogenic defects and, as a consequence, percutaneous procedures can be challenging due to unfavourable anatomical characteristics. To our knowledge, this is the first report on percutaneous complex VSD closure by using the two techniques described, i.e. the 'muscular anchoring' and the 'buddy wire delivery'.

The anchor-balloon technique is a well-established technique in the setting of percutaneous coronary intervention, and consists in a procedure in which a balloon is inflated distal to the target lesion to provide the anchor to deliver the interventional device.⁶ We believe that this technique should be used to provide further support to the guiding catheter, when advancing the guidewire through a tortuous VSD is challenging. The second technique, i.e. the buddy wire technique, is not entirely unique in the hole-closure business, as for example it was already described for atrial septal defect and ruptured sinus of Valsalva closure.^{7,8} An additional guidewire should be used in the setting of percutaneous VSD closure in order to give support while deploying the device,

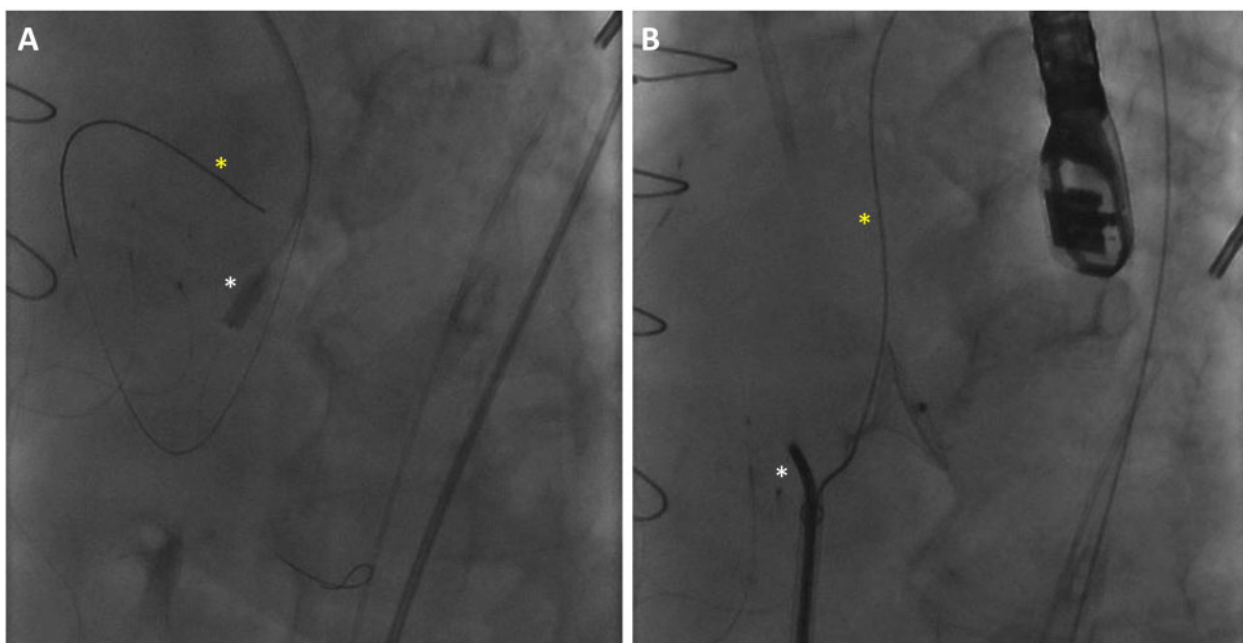


Figure 3 (A) The ‘muscular anchoring’ technique. A 4.0 × 8 mm non-compliant balloon (white asterisk) is inflated into the defect in order to provide support for advancing the exchange length guidewire in the pulmonary artery (yellow asterisk). (B) The Amplatzer Septal Occluder, still attached to the delivery cable (white asterisk), is positioned and deployed while stabilizing the system through the arteriovenous loop (yellow asterisk).

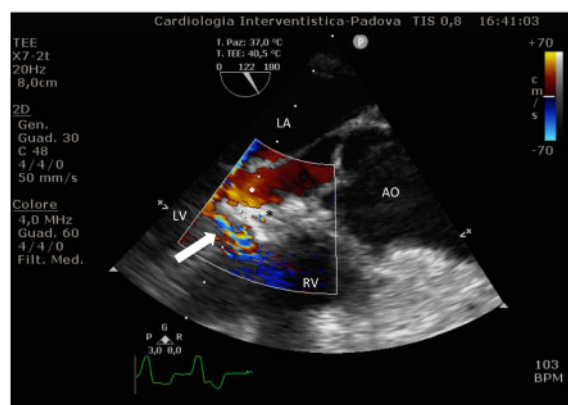
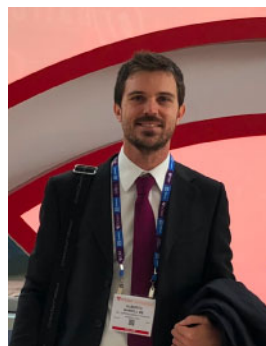


Figure 4 Post-procedural transoesophageal echocardiography showing minimal residual shunt. Note the Amplatzer Septal Occluder across the interventricular septum (asterisk) and the minimal residual shunt (arrow). AO, aorta; LA, left atrium; LV, left ventricle; RV, right ventricle.

and to maintain a useful access to the site even after the device sheath assembly has been withdrawn.

In conclusion, transcatheter closure represents an attractive minimally invasive approach to manage symptomatic iatrogenic VSDs. The techniques described, i.e. the ‘muscular anchoring’ and the ‘buddy wire delivery’, could help the operator to cross tortuous and tunnelled defects and to deploy closure devices in case of complex VSD anatomy, respectively.

Lead author biography



Alberto Barioli graduated in Medicine and Surgery at the University of Padua in 2011; he obtained a Postgraduate Diploma in Cardiology at the same University in 2017. He is the author of several articles published in national and international journals and of numerous presentations at national and international conferences on the topics of ischaemic and valvular heart disease. Since 2017, he works as a research fellow in the

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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: none declared.

References

1. Authors/Task Force members, Elliott PM, Anastakis A, Borger MA, Borggrefe M, Cecchi F, Charron P, Hagege AA, Lafont A, Limongelli G, Mahrholdt H, McKenna WJ, Mogensen J, Nihoyannopoulos P, Nistri S, Pieper PG, Pieske B, Rapezzi C, Rutten FH, Tillmanns C, Watkins H. 2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy: the Task Force for the Diagnosis and Management of Hypertrophic Cardiomyopathy of the European Society of Cardiology (ESC). *Eur Heart J* 2014;**35**:2733–2779.
2. Nishimura RA, Seggewiss H, Schaff HV. Hypertrophic obstructive cardiomyopathy: surgical myectomy and septal ablation. *Circ Res* 2017;**121**:771–783.
3. Okubo M, Benson LN, Nykanen D, Azakie A, Van Arsdell G, Coles J, Williams WG. Outcomes of intraoperative device closure of muscular ventricular septal defects. *Ann Thorac Surg* 2001;**72**:416–423.
4. Chessa M, Carminati M, Cao QL, Butera G, Giusti S, Bini RM, Hijazi ZM. Transcatheter closure of congenital and acquired muscular ventricular septal defects using the Amplatzer device. *J Invasive Cardiol* 2002;**14**:322–327.
5. Gersh BJ, Maron BJ, Bonow RO, Dearani JA, Fifer MA, Link MS, Naidu SS, Nishimura RA, Ommen SR, Rakowski H, Seidman CE, Towbin JA, Udelson JE, Yancy CW; American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines; American Association for Thoracic Surgery; American Society of Echocardiography; American Society of Nuclear Cardiology; Heart Failure Society of America; Heart Rhythm Society; Society for Cardiovascular Angiography and Interventions; Society of Thoracic Surgeons. 2011 ACCF/AHA guideline for the diagnosis and treatment of hypertrophic cardiomyopathy: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. *Circulation* 2011;**124**:e783–e831.
6. Fujita S, Tamai H, Kyo E, Kosuga K, Hata T, Okada M, Nakamura T, Tsuji T, Takeda S, Bin Hu F, Masunaga N, Motohara S, Uehata H. New technique for superior guiding catheter support during advancement of a balloon in coronary angioplasty: the anchor technique. *Catheter Cardiovasc Interv* 2003;**59**:482–488.
7. Haas NA, Soetemann DB, Ates I, Baspinar O, Ditkivskyy I, Duke C, Godart F, Lorber A, Oliveira E, Onorato E, Pac F, Promphan W, Riede FT, Roymanee S, Sabiniewicz R, Shebani SO, Sievert H, Tin D, Happel CM. Closure of secundum atrial septal defects by using the Occlutech occluder devices in more than 1300 patients: the IRFACODE project: a retrospective case series. *Catheter Cardiovasc Interv* 2016;**88**:571–581.
8. Awasthy N, Tomar M, Radhakrishnan S, Shrivastava S. Unconventional uses of septal occluder devices: our experience reviewed. *Indian Heart J* 2015;**67**: 128–135.