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Original Article

Percutaneous closure of moderate to large perimembranous ventricular septal defect in small children using left ventricular mid-cavity approach

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ABSTRACT

Aim & objective: To report mid-term follow-up result of transcatheter closure of perimembranous Ventricular septal defect (VSD) in children weighing less than 10 kg using Amplatzer Duct Occlude-I (ADO-I) by left ventricular (LV) mid-cavity approach.

Material & method: This is retrospective review of 35 children weighing less than 10 kg with moderate to large perimembranous VSD who were selected for transcatheter closure of VSD using ADO-I in between October 2016 to September 2018. Mean age was 2.08 ± 0.67 years (mean ± SD) and mean weight was 7.2 ± 1.2 kg (mean ± SD). Procedure was done by crossing the VSD from right ventricular side instead of using the standard approach by forming arterio-venous loop. Average fluoroscopic time was 9.2 ± 2.9 min (mean ± SD) and mean procedure time was 34.1 ± 13.1 min (mean ± SD). Mean follow-up period was 8.7 months (3–12 months)

Result: Device closure was successfully done in 32 patients. There was device induced aortic regurgitation (AR) in one case who was sent for surgery. One child was found to have complete heart block on first post-procedure day requiring permanent pace-maker implantation. During follow up none of them had any residual VSD, rhythm disturbance, AR and left or right ventricular outflow obstruction.

Conclusion: Device closure can be successfully done in moderate to large perimembranous VSD using left ventricular mid cavity approach in small children. LV mid-cavity approach has advantages in terms of lesser hemodynamic instability, lesser fluoroscopy and lesser chance of device induced AR than standard technique particularly in smaller children.

1. Background

Ventricular septal defect (VSD) is the most common form of congenital heart disease accounting for about 20% of all form of defects.[1] Until recently the standard treatment for VSD was open heart surgery. This is a major procedure that necessitates a sternotomy, cardiopulmonary bypass, blood transfusion, a permanent scar and potential risks of complete heart block, early and late arrhythmias, post pericardiotomy syndrome and even death.[2–6] Since the first report of VSD closure by trans-catheter approach by Lock et al in 1988,[7] the catheter based approach has now proven to be an alternative to surgery in many VSDs with acceptable mortality and morbidity as well as encouraging results.[8]

The scope of trans-catheter closure of moderate to large VSDs in small children is limited due to the larger size of delivery apparatus required which may cause hemodynamic instability and rhythm disturbance. So, surgical closure is the preferred option in this age group. Studies have shown that duct occluder can be safely used to close peri-membranous defects[9,10] by standard approach by forming an arterio-venous loop and deploying the device antegradely.[11,12]

In this series, we are reporting our immediate and short term results of VSD device closure in small children (weighing less than 10 kg) where Duct Occluders have been deployed using LV mid cavity approach. This technique has not been described in literature previously.

2. Materials and methods

This retrospective study was done in the Department of Pediatric Cardiology at Narayana Superspeciality Hospital, Howrah from

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October, 2016 to September 2018. Patients selected for VSD device closure and weighing less than 10 kg were included in this study.

2.1. Patient population

2.1.1. Inclusion criteria

(i) Children with moderate to large perimembranous VSD (hemodynamically significant VSD) with body weight less than 10 kg.

2.1.2. Exclusion criteria

(i) VSD closed by devices other than Amplatzer Duct Occluder-I (St. Jude Medical, St. Paul, Minnesota) (ADO-I) device, (ii) VSD other than perimembranous location, iii) Children < 10 kg with small restricted perimembranous VSD where heart failure symptoms were manageable by antifailure treatment, iv) Moderate to Large perimembranous VSD not amenable for device closure and sent for surgical closure

Total 35 patients who had fulfilled the inclusion criteria and underwent VSD device closure in between October 2016 to September 2018 using ADO-I device were included in this study. Pre intervention assessment of the size & the nature of VSD, suitability of device occlusion were assessed by TTE. Mean age of the study population was 2.08 ± 0.67 years (mean ± SD) and mean body weight was 7.2 ± 1.2 kg (Mean ± SD). Median size of the VSD was 6.1 mm (range 4.9–8.8 mm).

2.2. Device and technique

All patients were administered with 100 IU/kg heparin and antibiotics prophylaxis intravenously before the procedure. Procedures were performed under local anaesthesia and intravenous sedation.

We took 4Fr groin sheath in right femoral artery and 6Fr groin sheath in right femoral vein for all the patients. In each case, the VSD was profiled by left ventriculogram in standard 60° left-anterior oblique (LAO) with 20° cranially tilted view and 30° LAO, 30° cranially tilted views using a 4 Fr Pigtail catheter. Selection of the size of the device was based on echocardiographic and angiographic assessments (Fig. 1). We took ADO-I device 1–2 mm larger (RV end) than the maximum size of the defect measured on echocardiography. The VSD was crossed from the right ventricle (RV) by using a 4 Fr Judkin's right coronary artery catheter and 035″ J tip exchange length Terumo wire under fluoroscopic guidance in frontal projection (Fig. 2A), instead of crossing the VSD from the LV side as described in standard approach (by forming AV loop). After crossing the VSD from the RV side we parked the tip of the Terumo wire either in descending thoracic aorta (Fig. 2B) or in the left or in right subclavian artery. In all cases we exchanged the femoral venous groin sheath for ADO-I delivery system and crossed the VSD over the Terumo wire support. We used 6Fr delivery sheath for 8/6 and 10/8 mm ADO-I device and 7Fr delivery sheath for 12/10, 14/12 and 16/14 mm ADO-I device as per manufacturer's recommendation. We parked the tip of the sheath into mid-cavity of LV keeping the sheath across the defect (Fig. 2C).The desired sized ADO-I device was advanced through the delivery sheath and deployed under fluoroscopic guidance in the projection where VSD was profiled best (Fig. 2D).The final position of the device was assessed both by transthoracic echocardiography and by LV angiography (Fig. 3). Echocardiographic assessment included the alignment of the device with the ventricular septum, presence of significant residual shunt, impingement of aortic valve cusp by the LV disc of the device, presence of aortic regurgitation, any distortion in the coaptation mechanism of the aortic valve. Once found satisfactory the device was released. Our average fluoroscopic time was 9.2 ± 2.9 min (mean ± SD) and mean procedure time was 34.1 ± 13.1 min (mean ± SD).

2.3. Follow up

Tablet Aspirin was prescribed at a dose of 3–5 mg/kg/day to all the children after VSD device closure for 3 months. Clinical examination, electrocardiographic monitoring and TTE were performed on the day of discharge (first post intervention day), after 2 weeks, after 3 months and after 1 year of device closure.

2.4. Definition of complications

A major complication was defined as an event that resulted in death, long-term sequelae, need for immediate surgery, potentially life-threatening events, persistent arrhythmias needing pacemaker implantation, ongoing haemolysis requiring blood transfusion, thrombosis that required thrombolytic therapy, and increased valvular regurgitation needing device removal or drug therapy.

A minor complication was defined as an event that required drug therapy but was not life-threatening, with no long-term (>6 months) sequale, and which did not require long-term therapy. The following were also included in this group-haematoma of the groin, cardiac arrhythmias that required cardioversion or drug therapy during the procedure, minor degree reversible atrio-ventricular blocks, and transient loss of peripheral pulse needing only heparin therapy.

Procedural success was defined by device implantation in the appropriate position with no need for surgery/re-intervention (for example due to significant residual shunt or significant valve regurgitation).

3. Result

Ratio of pulmonary: systemic blood flow (Q_p: Q_s) was calculated in all the 35 patients. Median Qp: Qs was 3.8 (range 2.8–5.1). LV mid-cavity technique was performed in 33 cases and standard antegrade technique was performed in other 2 cases. We used ADO-I 8/6 mm device in 8 cases, 10/8 mm device in 20 cases and 12/10 mm device in 7 cases. Device placement using LV mid-cavity approach was successful in 32 cases. We could not cross the defect from RV side in 2 cases where the standard approach was followed using AV loop, of which device closure was successful in one patient and the other patient had significant AR as the LV disc of the device was impinging on aortic valve, prompting us to refer the patient for a surgical closure of VSD. In another child with perimembranous VSD with subaortic extension, though we followed the LV mid-cavity approach, there was significant AR as the device was catching the base of aortic valve cusp due to the close proximity of the defect with aortic valve. This patient was also referred for elective surgical closure of VSD.

In one child, we could not initially cross the defect from the RV side and we did the device closure using standard antegrade approach. However, there was development of severe AR due to impingement of the LV disc with aortic valve, which persisted inspite of multiple repositioning attempts. We retrieved the device, and in a repeat attempt we crossed the defect from the RV side and successfully deployed the device again as per our original plan. Interestingly due to better device alignment from this approach there was no AR and we could successfully release the device.

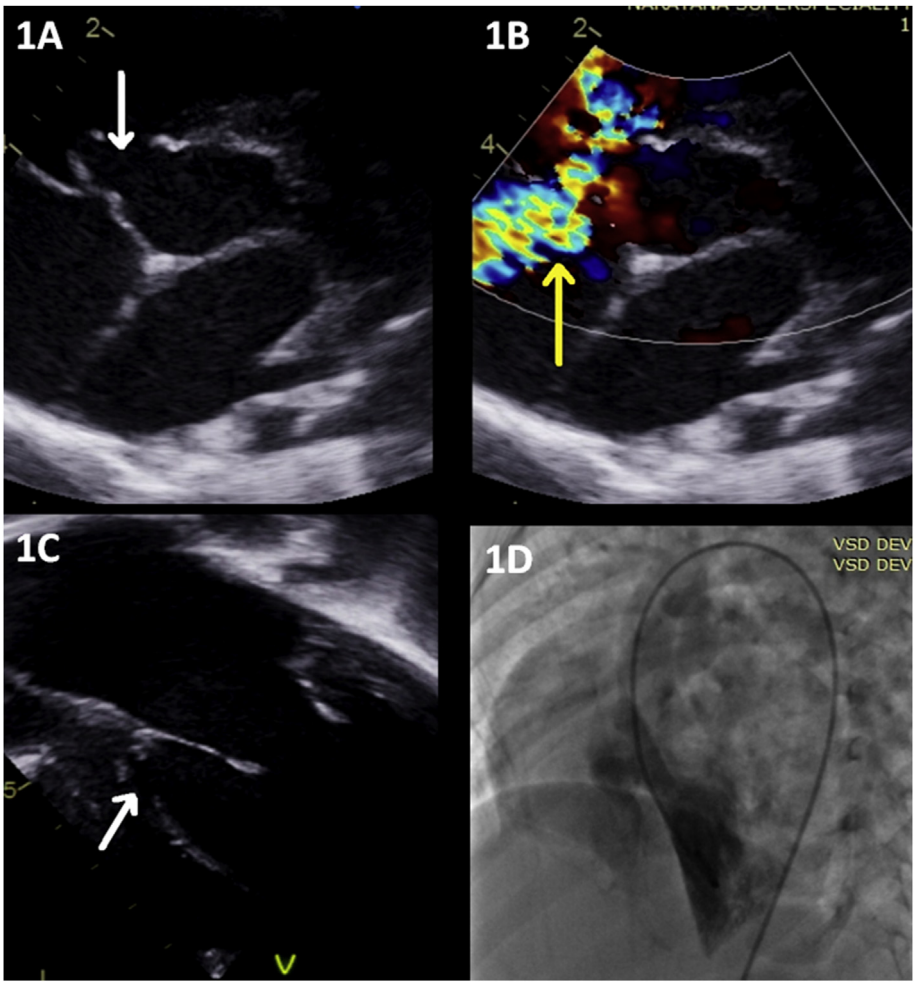

Fig. 1. Profiling the VSD- 2D echocardiography Parasternal short axis view showing large perimembranous VSD (White arrow) restricted by septal leaflet of tricuspid valve **(1A)**, Colour Doppler echocardiography showing large perimembranous VSD with left to right shunt with VSD jet directed towards right atrium as Tricuspid regurgitation (Yellow arrow) **(1B)**, 2D echocardiography Apical 4 chamber view showing moderate sized perimembranous VSD (White arrow) **(1C)**, Left ventriculogram (LV) in left anterior oblique view with cranial tilted projection showing moderate sized perimembranous VSD **(1D)**.

There was one immediate major complication. One child had developed complete heart block (CHB) on first post procedure day. We used 12/10 mm ADO-I device for that child. The child was started on intravenous Dexamethasone and kept under observation for 72 h. Surgical removal of device and surgical closure of VSD was done on 4th post procedure day as the child continued to have CHB. Rhythm reverted to sinus for few hours after the surgery and subsequently the child continued to have CHB and permanent pacemaker was implanted on 4th post-operative day. Four children had transient loss of femoral pulse, which was managed successfully with 24 h of heparin infusion.

Mean follow up period of this study was 8.7 months (3–12 month). During follow-up, 9 patients had trivial intra-device residual shunt on day 1 of procedure. None of them had any residual shunt neither any left or right ventricular outflow obstruction at 3 month follow-up. All of them had good weight gain during follow-up. There was significant reduction of LV dimension during 3month follow-up with normalisation of Z score of LV dimension in diastole in all of them where device closure was successfully done. None of them had any arrhythmia (apart from one child who had CHB requiring pacemaker implantation), haemolysis, thromboembolic event or endocarditis on follow-up.

4. Discussion

Moderate to large VSDs should be closed early because of its hemodynamic significance (heart failure, early development of pulmonary vascular obliterative disease) and as the chance of spontaneous closure is negligible. Traditionally, surgery is the treatment of choice for large VSD in smaller children, but it does have some potential risks of complications including CHB in 1–5% of the cases,[2,13,5,6] significant residual VSD in 1–10% of the subjects,[13-17] the necessity for re-operation in 2% of the patients,[11] and even death in 0.6–5% of the cases.[13–16] Furthermore, infections, tachyarrhythmias, and neurological complications may occur after surgery.[13]

Considering the fact that trans-catheter closure of VSD is less invasive than surgery with a relatively lesser period of hospital stay, we have attempted to do trans-catheter closure for these patients. The intermediate term follow up study has shown good outcome with a success rate of 88.6%.

In our experience, proper patient selection is the most important factor for procedural success. For successful VSD device closure, pre intervention profiling of VSD by TTE and intraoperative profiling during LV angiography are extremely important. We have stressed on the measurement of the size of the defect, it's location

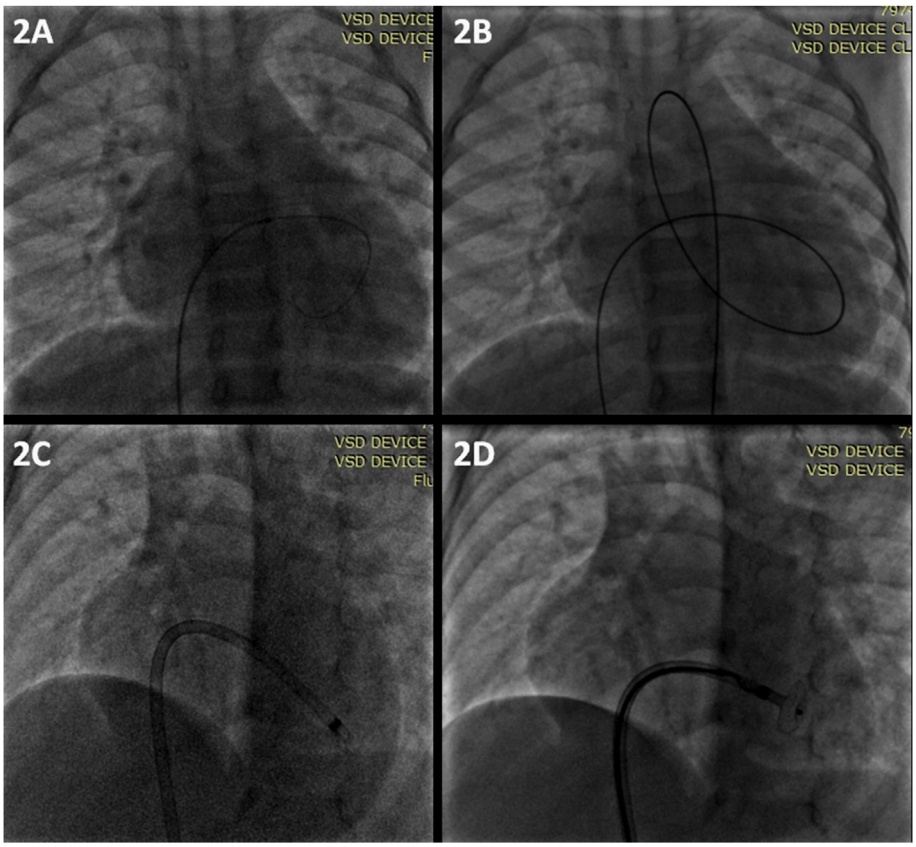

Fig. 2. VSD closure using LV mid cavity approach- VSD being crossed from right ventricular side in frontal projection using a J tipped Terumo wire **(2A)**, Tip of the wire being parked in descending thoracic aorta after crossing the VSD from right ventricular side **(2B)**, Device delivery sheath being parked at LV mid cavity **(2C)**, VSD device being deployed in left anterior oblique view with cranially tilted projection **(2D)**.

and extension of the defect into the inlet, trabecular or outlet septum and distance of the defect from the aortic valve on TTE. Length of subaortic rim and the distance from the aortic valve from the upper edge of the defect was measured and the length 4 mm or more was considered to be adequate. We measured the defect in orthogonal views (apical four chamber view, parasternal long and short axis view and subcostal view) (Fig. 1) and took the maximum measurement as the working size-based on which size of the device was selected. We have oversized the defect by 1–2 mm (RV end) in all our cases, so that the device fits snugly, minimising the chances of migration (we have used 10/8 ADO-I device for 6–7 mm defect).

We have used LV mid-cavity approach instead of the standard antegrade approach (using A-V loop).In our experience, the standard approach, if attempted in a small heart, leads to tremendous hemodynamic compromise because of splinting of heart by the delivery sheath passing into the ascending aorta across the VSD through the tricuspid valve and RV. The splinting is also evident during snaring for arteriovenous loop formation and also if a stiff wire is used as a support for the delivery sheath in a relatively smaller heart.

While deploying the device from the aorta using standard antegrade approach, we have often seen that there is increased chance of the LV disc of the device impinging on aortic valve resulting in formation of AR. Aortic valve being an anterior structure than the perimembranous area comes in between the LV disc of the device and perimembranous defect while bringing the device from aorta. This complication is also much more common in smaller children particularly while putting a relatively larger device. In LV mid-cavity approach chance of hemodynamic compromise is much less as the delivery sheath is being placed in LV cavity after crossing the VSD with less chance of splinting of heart against the sheath. LV cavity being a posterior structure than perimembranous area of septum and aortic valve being the anterior structure, chance of LV disc of the device impinging on the aortic valve is less in LV mid-cavity approach when the aortic rim of the defect is adequate. This was very well highlighted in our patient who had significant AR by standard approach, with complete disappearance of AR when the device was deployed from LV mid-cavity approach.

Crossing the defect from RV side is a challenge and it is pertinent that the operator should be well versed with the general nuances of the cardiac anatomy. We used 4Fr Judkin's right coronary artery catheter and 035 exchange length J-tip Terumo wire for crossing the defect. We crossed the defect using fluoroscopy in frontal projection. After parking the catheter in the pulmonary artery it is gently brought down with a slight clockwise rotation to face the tip posteriorly towards the perimembranous septum. Once near the left para-spinal area at approximately the central point of the cardiac silhouette the wire can usually be passed into LV across the VSD. We ensured that the J wire formed a loop in the LV and then advanced it into descending aorta or either subclavian arteries. This provides enough support for the 6 Fr/7 Fr sheath to pass into LV thus obviating the need for a stiff wire in a small heart (Fig. 2).

One patient developed CHB which accounts for the incidence rate of 2.8% in this case series. This is comparable with the rate of CHB following surgical closure of VSD published in various literatures.[2,5,6] The child had undergone surgical removal of device and surgical closure of VSD after which rhythm reverted to sinus for few hours and again developed CHB requiring permanent pacemaker implantation. Intraoperatively, surgeon found the defect to be located in the inlet perimembranous septum and the position of the

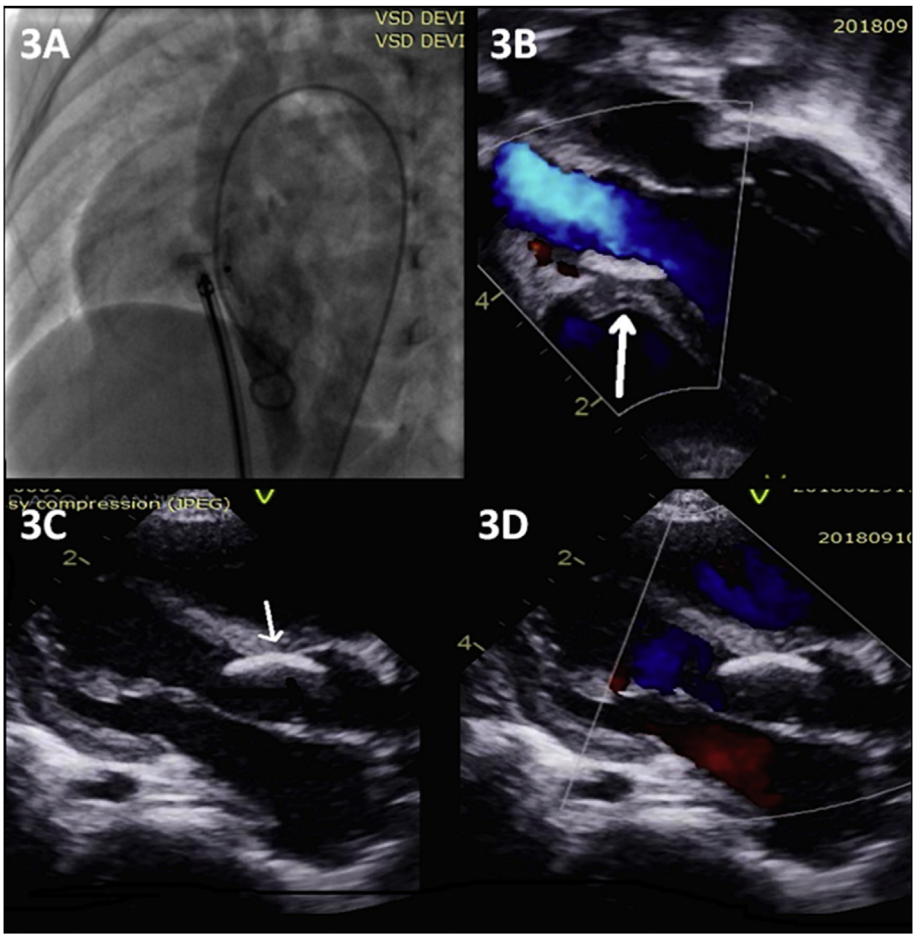

Fig. 3. Post device closure assessment- LV angiogram showing stable device position away from aortic valve without any significant residual **(3A)**, Color Doppler echocardiography Apical five chamber view in systole showing device in stable position (white arrow) without causing any LV outflow obstruction **(3B)**, 2D echocardiography Parasternal long axis view showing device (White arrow) away from aortic valve without hampering it's coaptation (Black arrow) **(3C)**, Color Doppler echocardiography Parasternal long axis view in diastole showing device not causing any aortic regurgitation**(3D)**.

device was very stable. The defect was big, and the device looked bulkier compared to the size of the heart though it was not oversized compared to the defect. This patient had a relatively large defect requiring 12/10 mm ADO-I device, but 7 patients in this case series required a device similar or bigger than 12/10 mm ADO-I. So, device size may not be the only determining factor for development of CHB. Rhythm abnormality persisted after surgical removal of device and surgical closure of VSD. This child may have had the conduction bundle in extreme close proximity to the defect which got affected by the shearing force exerted by the disc of the device and also by the surgically placed patch. Development of CHB is unpredictable and to some extent unavoidable complication of VSD device closure particularly in small children.

In this study we have shown that the fluoroscopy time and total procedure time is relatively short in LV mid-cavity approach and there are lesser chance of hemodynamic compromise and development of device induced aortic regurgitation in comparison to standard approach particularly in small children.

4.1. Study limitations

Our study was done at a single centre. Also the study population was small and the follow-up duration was less. The results of this study should be supported by a multicentre study involving many operators with a larger number of patients.

5. Conclusion

Percutaneous closure of moderate to large perimembranous ventricular septal defects can be successfully done in children weighing less than 10 kg and LV mid-cavity approach is the preferred method than the standard antegrade technique. There are lesser chances of hemodynamic compromise and development of device induced aortic regurgitation. Also, the fluoroscopy and total procedure times are lesser in device closure using LV mid-cavity approach.

Conflicts of interest

All authors have none to declare.

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