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Case Report

Transcatheter self-expanding stent closure of sinus venosus atrial septal defects with indigenous method of determining appropriate stent length without advanced imaging techniques -A case series of single centre experience

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A B S T R A C T

Transcatheter closure of sinus venous Atrial Septal Defect (SVASD) was proving to be safe and effective with accumulating evidence. We report 3 cases of successful transcatheter closure of SVASD with indigenous reproducible technique without need for advanced imagining techniques with favourable outcomes. Three patients aged 27,42 and 44 years underwent successful closure of SVASD using self-expandable Endurant™ II stent graft (Medtronic) after undergoing pre-procedure successful balloon occlusion of Superior Vena Cava (SVC) without obstructing right upper pulmonary vein (RUPV) drainage. We used fluoroscopy, *Trans*-esophageal echocardiography (TEE) and marker pigtail for guiding measurements to choose appropriate stent size. One patient developed cardiac tamponade requiring surgical removal of stent with surgical closure of the defect.
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1. Introduction

The sinus venosus atrial septal defect (SVASD) first described by Peacock in 1858 and it represents 5–10% of atrial septal defects (ASD).[1] Surgical closure of the defect remained the treatment of choice until a novel transcatheter method of sealing the defect with balloon expandable stent in 2014 by Garg et al.[2] Currently transcatheter closure is being used as an alternative to surgical repair and the results are encouraging. Since this procedure is technically demanding and the complication rates are too frequent, there has been a lot of modifications in the original technique by individual operators across the globe. We describe one such technique to close these defects with the available tools in the cardiac catherization lab.

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2. Case series

2.1. Pre procedure evaluation

All the patients underwent clinical examination, Blood investigations, CXR (chest x ray) and ECG(Electrocardiogram). 2D & 3Dimensional TEE was done to delineate the anatomy of the defect. Defects extending beyond cavoatrial junction were excluded. Patient selection for stent closure was based on test balloon occlusion of SVC with unobstructed flow of RUPV to LA (Left Atrium) during evaluation.

2.2. Access and septal puncture

All patients received IV cefuroxime(25mg/kg) before initiation of the procedure. The procedure was done under general anaesthesia. Both femoral venous accesses were taken, and 6 Fr Terumo valved short sheaths were placed. Invasive femoral arterial blood pressure monitoring was done. As Patent foramen ovale (PFO)was present in 2 cases, hence *trans*-septal puncture of fossa ovalis was

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required in one patient. IV heparin 100 IU/Kg was administered after *trans*-septal puncture. Using a 5Fr pigtail catheter pulmonary artery angiogram was done and return of contrast through the RUPV into RA (Right atrium) during levophase was confirmed. A 5 Fr Judkins catheter (Cordis, Milpitas, CA, USA) was placed through the PFO in the RUPV to check the flow and pressure monitoring during balloon occlusion test.

2.3. Balloon occlusion of SVC

Prior to stenting cases were assessed for unobstructed RUPV drainage after balloon occlusion of SVC. RFV (Right femoral vein) sheath was upgraded to 18 F short sheath (Cook medical, USA). An Amplatzer super stiff wire (Boston Scientific Corporation, Natick, MA) was passed through the multipurpose catheter and was placed in the right SVC. A 30mm $\times$ 50 mm Z - Med Balloon (B. Braun Med Inc, Germany) was taken over the stiff wire and was positioned across the defect. The balloon was inflated so that it completely covers the upper end of RUPV-SVC junction and the lower end of balloon completely occludes the defect (Fig. 1C). Then Selective RUPV injection was done using 5F Judkins catheter which was placed earlier (Fig. 1 B, C). Unobstructed RUPV flow into LA without raise in pressure along with complete occlusion of the defect was confirmed and then it was decided to proceed for transcatheter closure of the defect. Contrast ECHO was also done during balloon inflation with the catheter in RUPV using agitated saline showed RUPV draining into LA without any obstruction and no residual shunt.

2.4. Drawing landmarks for determining stent length and diameter

After assessment of balloon occlusion of SVC, Fluoroscopic cine images and TEE was used to decide the length of the stent. Fluoroscopy images done in Anteroposterior (AP) projection were used for measurements and table is frozen once we started the calculation/implantation procedure. Selective RUPV and Left innominate vein/SVC injections were done to draw the landmarks (Fig. 1 A). The SVC injection was done, and maximum diameter of SVC was measured at the SVC -RA junction. The maximum diameter between the lateral wall of RA and the atrial septum just below the defect was measured by TEE. We added 4mm to this diameter to determine the stent diameter. A 5F marker pigtail catheter was placed in SVC and another 5Fr pigtail was advanced into the SVC which was simultaneously visualized in TEE as well as in fluoroscopy. The pigtail was withdrawn slowly visualizing it in both fluoroscopy and TEE (Fig. 3A). The point at which the head of pigtail crosses the Left innominate vein (LIV) and SVC junction (approximately below the drainage of azygous vein), marked as proximal covered portion part of stent (Fig. 2 D). Pigtail was further withdrawn, and as it crosses the lower margin of the defect, we marked 5mm below the pigtail head on the fluoroscopy screen to make it as desired distal end of the stent (Fig. 2 D).

2.5. Placement of stent

RFV sheath was further upgraded to 20 Fr short sheath (Cook medical, USA). A Multipurpose catheter using 0.035-inch Terumo wire (Terumo Corporation, Tokyo, Japan) was parked in SVC. Terumo wire was exchanged for an Amplatzer super stiff wire. Over the stiff wire, the desired Endurant II™ stent (Medtronic) graft system was taken and positioned with previous fluoroscopy marker and TEE guidance. Stent was Placed with check angiograms to reconfirm the position. We slowly deployed the stent and made sure that lower portion coincides with inferior margin of the defect we drew on the fluoroscopy screen. Post stent deployment RUPV angiogram was done to show unobstructed flow into LA without any residual shunts (Fig. 2 F). The RUPV pressure is also recorded to rule out obstruction. A TEE doppler & bubble contrast echo was done in RUPV to confirm residual shunt (Fig. 3 B).

3. Case details

All cases had successful SVASD stenting. Age of the patients were 27, 42& 44Yrs respectively (as shown in Table 1). All had symptoms of NYHA II initially which had progressed to NYHA III respectively at the time of diagnosis. Patient 3# is a known

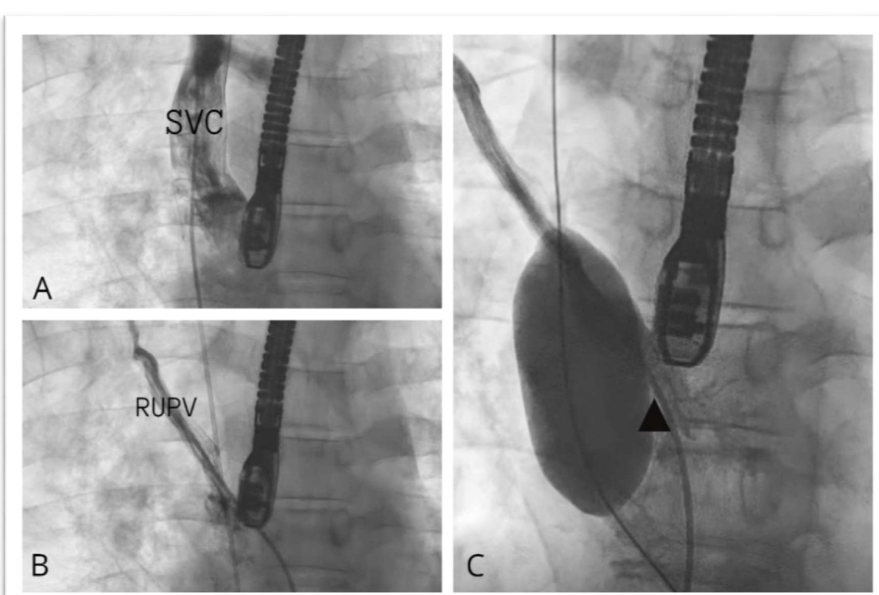

Fig. 1. Angiographic images of Superior vena cava(SVC) and Selective Right upper pulmonary vein(RUPV) angiography showing draining of RUPV into right atrium through atrial septal defect(A,B). Balloon occlusion of SVC and selective pulmonary angiography showing unobstructed drainage of RUPV into left atrium (Dark arrow head in C).

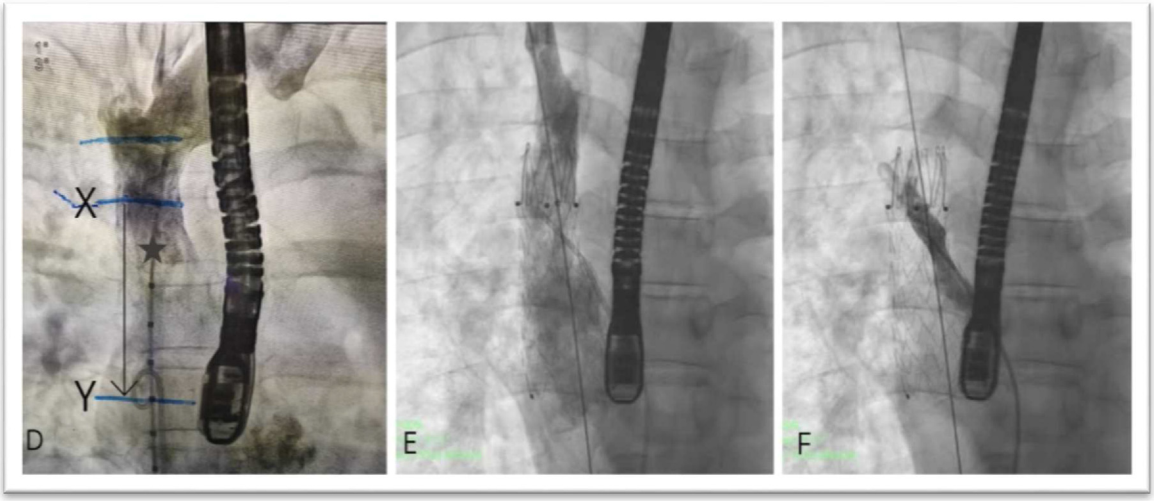

Fig. 2. Marker pigtail (solid star) & Normal pigtail simultaneously viewed in fluoroscopy still image after visualizing it in the *trans*-esophageal echocardiography (TEE) and a line is drawn at the beginning of curvature below the pigtail head(Y) kept at lower margin of the defect and it is marked on fluoroscopic still image as the lower end of covered portion of stent. Same pigtail pushed up and line is drawn below the drainage of azygous vein(T3-T4) in the still fluoroscopic as well as TEE image as upper margin of covered portion of stent (X). The distance between points X &Y is the covered portion of the stent(D).(E&F) successful deployment of stent graft and selective right upper pulmonary venography showing unobstructed drainage into the left atrium without any residual shunt across the defect.

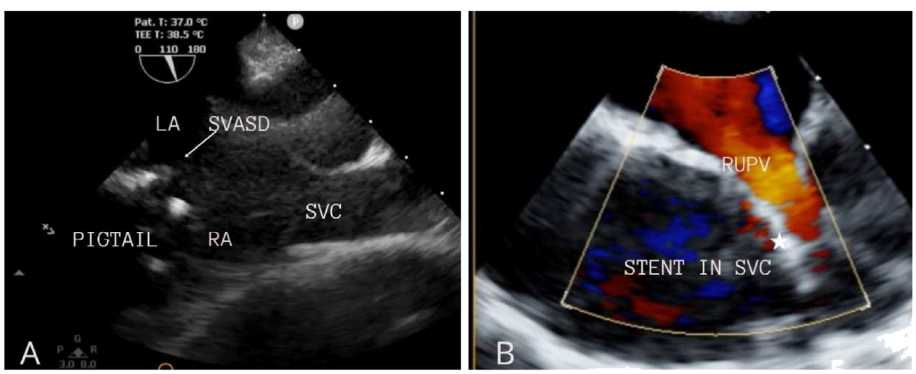

Fig. 3. A. Transesophageal Echocardiogram Showing pigtail in the lower end of sinus venous atrial septal defect (SVASD). Successful deployment of stent creating partition (solid star) between SVC and Right upper pulmonary vein (RUPV) with unobstructed drainage of RUPV into Left atrium (3 B).

Table 1
Characteristics of patients and their outcome after stenting.

Characteristics	Case ≠1	Case≠ 2	Case ≠3
Age	27years	42 years	44years
Sex	Male	Female	Male
weight	65kg	79kg	83kg
Qp: Qs	2.3:1	3:1	2.5:1
RUPV access	PFO	PFO	Transeptal puncture through femoral venous approach
Balloon occlusion Test	successful	successful	Successful
Stent length (Covered portion)	70 mm	49mm	70 mm
Stent Diameter	36mm	32mm	36mm
Residual shunt	Nil	Nil	Nil
Complications	Nil	Hemopericardium after 24 hours	Transient atrial ectopics

hypertensive and Type II diabetes mellitus on treatment. He also underwent coronary angiogram prior to procedure, which was normal. All three patients never had complications during procedure and had successful stenting. No residuals or pulmonary vein compression was noted after-stent deployment. Heparin infusion at 10 IU/Kg overnight after the procedure was started along with dual antiplatelet cover (Aspirin & clopidogrel 75mg each) which was advised to continue for 6 months. Patient ≠1 & ≠3 came for post procedure follow up at 1 month. On follow up TTE showed stent in position with good flow and there was no residual shunt. Patient ≠ 2 underwent successful transcatheter closure of SVASD defect. She was extubated immediately post procedure and was observed in recovery. Surveillance TTE done prior to transfer out from recovery room after 4 hours showed stent in position, no residual shunt with no pericardial effusion. Later in the evening she developed hemodynamic compromise with acute onset interscapular area pain.

Repeat TTE showed pericardial effusion with signs of cardiac tamponade. Emergency pericardiocentesis showed hemorrhagic fluid with transient improvement in vitals. Pig-tail catheter was left in situ and she was monitored in CICU (Cardiac intensive care unit), with minimal ionotropic support. Repeat TTE showed recurrent pericardial collection despite continuous drainage and her hemodynamics later started to deteriorate. Hence, she was taken for emergency exploratory thoracotomy. Intrapericardial exploration revealed stent anchoring pins/barbs protruding out through SVC, scraping the adjacent ascending aortic wall with a tiny rent in the ascending aorta. Surgical removal of stent and repair of rent in ascending aorta along with closure of SV ASD was done. After successful surgical correction she was stable and discharged on post-operative day 5. She is doing well on her first follow up with no residual shunt with sinus rhythm. Stenting patients follow up showed regression of the dilated RA &RV with normal sinus rhythm in ECG without any residual shunt.

4. Discussion

Surgical closure of SVASD is associated with incidence of arrhythmia, residual shunts, or patch dehiscence post-surgery.[3] Compared to transcatheter closure still surgery is a cost-effective attractive option, however effects of cardiopulmonary bypass, blood products use, and intensive care stay must be considered. Recently non-surgical closure of sinus venous ASD is gaining importance in the field of interventional cardiology due to better understanding of anatomy and advancement in the imaging techniques.[4–6] Various modification of the original transcatheter technique by Garg et al.[2] was made according to the resource availability in developing countries without the need for advanced imaging and 3D printed model stimulators. Most of the cases after assessment by TEE were deemed fit for transcatheter closure in adults unless the defect is high up in the SVC. We described one such technique which is easily reproducible with high success rate in every catherization lab equipped with TEE. Pre procedure imaging is required to get the desired stent length and so to avoid stent fore shortening or embolization post deployment. In developing countries as a cost cutting measure and to prevent use of multiple stents, we considered using single stent with appropriate length will be effective and feasible than balloon expandable stent. In our cases we used Endurant™ II stent graft (Medtronic) which was used for endovascular aneurysm repair (EVAR) with high success rate,[7] so we used as on off label basis to close SVASD.

Further Challenges for use of Balloon expandable stent are:

- Use of 10-zig covered stent for larger diameter SVASD defects were not widely available in market.
- These covered stents don't have anchoring pins to prevent embolization and migration of stents.
- Higher chance of using multiple stents, when a foreshortening occurs after deployment of covered balloon expandable stents.[8]

Advantages of self-expanding stents:

- Single stent is enough with proper determination of required stent length.
- Anchoring pins also reduces the risk of embolization after deployment with better flexibility.

Abdullah et al.[9] showed long term follow up of stenting of SVASD showed good outcome without any serious complication except for residual shunt which was later closed with device and covered overlapping stents. We postulated two possibilities for the onset of pericardial effusion in our patient ≠ 2, one possible cause is that those anchoring pins protruding out of SVC would have caused tiny rent in the aortic wall near the SVC would have led to hemopericardium and other reasons could be the slow expansion of the stent graft after deployment could have led to projection of those barbs out of SVC and causing injury to aorta. Most common complications after stenting are stent migration, thrombosis, and misplacement.[8] Choosing appropriate stent length is utmost important once balloon interrogation is over. Use of stents with barbs in our cases gave additional advantage of preventing stent misplacement and migration. Using pigtail near the lower end of the defect in a cine fluoroscopy image to measure the required stent length was encouraging to avoid oversizing and at the same time covering the entire defect was an added advantage. Success of structural cardiology intervention depends on preprocedural imaging and knowing the anatomical variations. Computerized Tomography (CT) guided sizing would have given an edge in understanding morphological variations such as landing zones for the anchoring pins of the stent and to delineate the distance between SVC & ascending aorta and cardiac tamponade could have been prevented. We also propose that simultaneous angiogram of SVC and aortogram should be done to look for adequate separation of the SVC and ascending aorta for all the cases before stent placement and to determine the zone in SVC to place the upper end of the stent, so that we can avoid anchoring pins encountering the aortic wall and thereby preventing serious pericardial effusion and vessel injury.

5. Conclusion

Successful transcatheter closure of sinus venosus ASD with self-expanding stents can be done but there is likely chance of barbs causing vessel injury if the partition between SVC and Aorta is inadequate.

Conflict of interest

None.

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None.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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