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Transcatheter Sinus Venosus ASD Closure: A Case Series of First Ever Transcatheter Closure of the Defect in Pakistan

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Abstract

Superior sinus venosus atrial septal defect is a direct communication between the right and left atrium located above the fossa ovalis below the junction of the superior vena cava and the right atrium. It is associated with partial anomalous pulmonary venous drainage, especially of the right upper and middle pulmonary veins. Over the years, surgical repair of the defect has been primary approach to close that defect. However, since 2014, percutaneous closure of the defect has gradually become a safe and effective alternative to surgery in carefully selected patients, although worldwide experience remains limited. This case series highlights three cases of first ever transcatheter sinus venosus atrial septal defect closures in Pakistan.

Understanding the anatomy of the crossroads formed by the Right Upper Pulmonary Vein (RUPV) ostium, the SVC orifice, the Right Atrium (RA) and the Left Atrium (LA) above the level of the unroofed atrial septum is key to appreciating the procedural concept. This article provides a comprehensive, up-to-date description of percutaneous management of SVD, detailing preprocedural screening, multimodality imaging for case planning and guidance and procedural steps. It also summarizes a comprehensive overview of the contemporary clinical outcomes of the transcatheter SVD closure. It also underscores the importance of the fact that transcatheter closure of this defect can easily be achieved, significantly reducing patient's morbidity as opposed to open heart surgery.

We believe that modified techniques along with growing experience of interventionalists all over the world will further enhance safety and expand the role of transcatheter closure of SVD.

Keywords: Anomalous venous drainage; Interventionalists; Transcatheter; Sinus venosus

Introduction

Sinus venosus atrial septal defect is a congenital anomaly which results from abnormal communication between both the atria and is always associated with partial anomalous pulmonary venous return. This defect is surgically closed using patch closure and relocating the pulmonary veins to the Left Atrium (LA). Transcatheter closure has been achieved through placement of a covered stent in the Superior Vena Cava (SVC) [1]. In 2013, Abdullah et al, described an innovative technique successfully used in 4 patients, where covered balloon-expandable stents were deployed across the superior cava–right atrial junction, replacing the deficient posterior wall and redirecting flow from the right pulmonary veins (RPVs) to the left atrium [2]. Subsequently, there have been several case reports and small series describing the application of this approach [3-5]. We report first ever transcatheter sinus venosus atrial septal defect closure in the country. These novel procedures were performed with the assistance of a foreign proctor at the Armed forces Institute of Cardiology, Rawalpindi, Pakistan.

Case Series

Case no: 1

Case history and examination

The first case was a 30-year-old male. He presented with left sided chest pain along with exertional dyspnea. He underwent 2D as well as transthoracic echocardiography which showed sinus venosus ASD with the right upper pulmonary veins draining into the SVC-RA junction while the rest of the veins draining

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normally into the left atrium. His CATH data showed a Qp-Qs 1.5:1 and PVR 1.4 wum2. CT angiography was done which revealed sinus venosus defect with the anomalous venous drainage **(Figure 1)**.

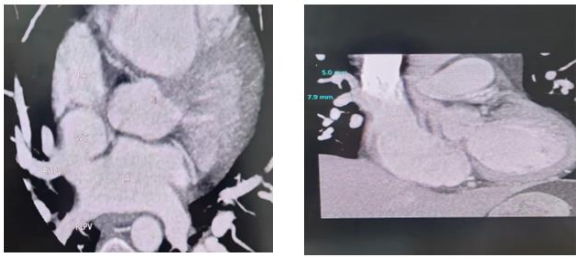

Figure 1: CT Angio axial and oblique images show sinus venosus defect as well as anomalous venous drainage.

Methods

The procedure was done under fluoroscopic and transesophageal guidance. Three access lines were taken namely the right femoral vein, left femoral vein and the right internal jugular vein. A 150 cm Terumo wire was introduced from the right femoral vein and kept into svc as guide for jugular venous approach. Terumo wire was then replaced with a 0.035 cm, 260cm, super stiff wire.

This wire was then snared with a 5F snare introduced from the right internal jugular vein. A 22 x 40mm VACS III balloon was introduced from the right femoral vein and inflated across the defect for sizing. The position was further confirmed with a transesophageal echo. A 5F JR was placed into the right upper pulmonary vein to look for rise in pressures post occlusion. Pre occlusion pressures were 7/4/3 mmHg. An 80mm Optimus XXL PTFE covered stent was selected. A 12 x 60 mm OTW mustang balloon was introduced through the right femoral vein and exteriorized from the internal jugular vein.

A 100cm long surgical Mersilk suture with a straight needle was passed through one of the upper struts of the stent with the intent to apply traction to the stent. The stent was then crimped onto mustang balloon and introduced from the internal jugular vein. The 12 mm balloon was removed and the stent was then partially inflated with a 26 x 40 mm VACS II balloon. Afterwards the caudal portion of the stent was flared with a 28 x 30 mm VACS II balloon. **Figure 2** shows the stent deployed at the SVC-RA junction. The suture was removed and venogram was done again which showed diversion of anomalous pulmonary venous drainage into the left atrium. This was again confirmed with a transesophageal echo. Pressures in the diverted vein post occlusion of the defect remained the same.

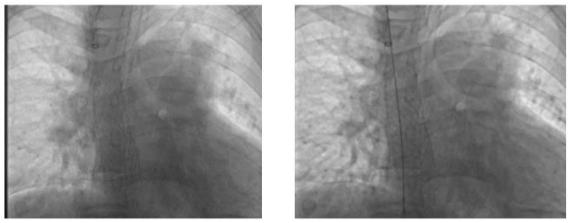

Figure 2: Cath images showing the stent at the SVC-RA junction.

Case no 2

Case history and examination

The second case was a 7-year-old boy who presented with shortness of breath on exertion (NYHA II). He too underwent 2D echocardiography followed by CT angiography which revealed a large sinus venosus atrial septal defect measuring 18mm with right upper pulmonary veins draining into the SVC-RA junction **(Figure 3)**.

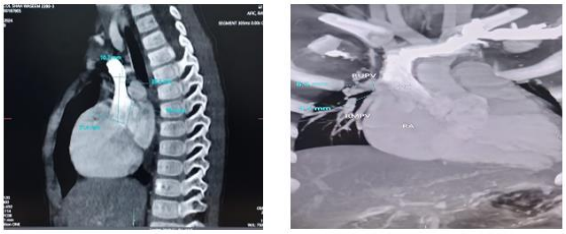

Figure 3: CT angio sagittal and coronal views showing the defect as well as anomalous venous drainage.

Methods

Three access lines were taken namely the right femoral vein, the left femoral vein and the right internal jugular vein. Terumo wire was passed from the right femoral vein and kept in svc as a guide for accessing the internal jugular vein. An SVC angiogram was done which showed the position of the innominate vein. A 5fr JR was kept in the right upper pulmonary to note the pressures pre and post occlusion of the defect. The pressures pre occlusion were 7-3-5 mmHg. A 0.035 260cm super stiff wire was introduced from the right femoral vein and exteriorized from the right internal jugular vein. An 18 x 30 mm VACS II balloon was used to size the defect. A 12 x 50 mm Altosa balloon was passed from the right femoral vein and again exteriorized from the right internal jugular vein. A 57mm Optimus XXL stent was select to be placed across the defect and a 100 cm long suture with straight needle was passed through one of the struts of struts of the stent. The stent was then crimped onto the balloon and introduced from the internal jugular vein. Cranial end of the stent was expanded using a 12 x 50 mm balloon while the caudal end was flared with a 24 x 40 mm VACS II balloon **(Figure 4)**. Finally, the diversion of the anomalous

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pulmonary veins into LA was confirmed by angiogram as well as transesophageal echo. The pressure in the right upper pulmonary vein post occlusion was 8-3-5 mmHg.

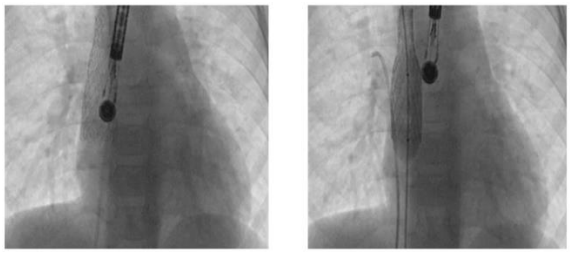

Figure 4: Cath images showing the balloon inflation of the stent. 5F JR placed in the anomalous vein.

Case 3

Case history and examination

The last case was a 13-year-old girl who had presented with complaints of chest pain and palpitations. She too underwent 2d echocardiography which revealed dilated right atrium and ventricle. Sinus venosus defect was not picked on the transthoracic echo. Hence, she underwent cardiac cath which revealed anomalous right upper pulmonary veins draining into SVC-RA junction with Qp Qs 2:1 and PVR<1 wum2. Her CT angiography showed sinus venosus atrial septal defect with anomalous right upper pulmonary veins **(Figure 5)**.

Figure 5: CT angio sagittal and coronal images.

Methods

Three access lines were taken namely the right femoral vein, left femoral vein and the right internal jugular vein. A 5F JR was introduced from the left femoral vein and placed into the anomalous right sided veins to look for changes in pressure after occlusion of the defect. The baseline pressures noted were 7-3-4 mmHg. An SVC angiogram was performed to look at the position of the innominate vein. A 260 cm super stiff wire was introduced from the right femoral vein with the help of a 5F Judkins Right catheter and exteriorized from the internal jugular vein.

The initially placed 7F short sheath in the internal jugular vein was replaced with a 14F dry seal. The sizing of SVC was done using a 18 x 30 mm VACS II balloon. A 57 mm XXL Optimus PTFE covered stent was selected and it was crimped onto 12 x 50 mm Altosa balloon. The stent with the balloon was introduced from the right internal jugular vein into svc. Similar to previous cases, a 100 cm long Mersilk suture with a straight needle was passed through one of the struts of the stent with the intent to anchor the stent and to provide traction from the cranial end for proper positioning. The caudal end of the stent was then ballooned with a 24 x 50 mm VACS II balloon.

Following deployment, the stent migrated downwards into the RA leaving the defect open. Hence, another 43 mm covered stent was used to cover the cranial end of the defect. Again, the cranial end of the defect was left uncovered so another 48 mm Optimus XXL stent was deployed. Initially, the angiogram along with transesophageal echo showed adequate closure of the defect but after a short while the combined assembly of all the three stents migrated downwards into the right atrium. Finally, an 80 mm stent was deployed to cover the defect as well as to provide support to the previously placed stents **(Figure 6)**. The transthoracic echo showed complete obliteration of the defect and the angiogram showed the same. Right pulmonary venous pressure post occlusion was 7-5-3 mmHg.

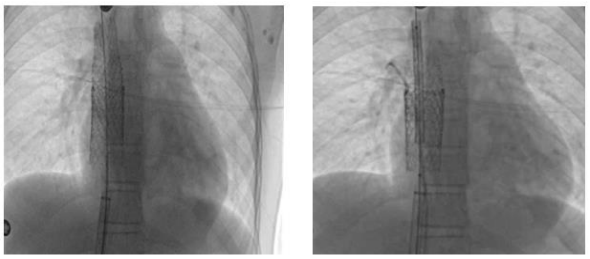

Figure 6: Cath images showing stent within stent.

Discussion

We report the first ever transcatheter closure of the sinus venosus defect in the country. The first case of sinus venosus ASD percutaneous correction was published in 2014 by Garg et al. and has since been performed in several countries. A number of cases have been reported to date [6,7]. This approach involves placing a covered stent from the SVC through the internal jugular vein into the right atrium leading to diversion of the anomalous pulmonary veins to the left atrium through the posterior part of the ASD; the anomalous vein is thus excluded from the upper systemic return. Therefore, this is not precisely an ASD closure, but more of a blood flow redirection. We believe this technique could have been inspired by a surgical procedure.

The technique used here was a modified version of the previously used transcatheter techniques. This

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technique was devised by Dr Younes B. It involves the usage of the surgical Mersilk suture which helps in providing traction to the stent from the SVC and is helpful in positioning the stent.

Conclusion and Results

Post procedure 2d echocardiography was done in all the patients which showed the stent at the SVC-RA junction with the pulmonary veins draining into the left atrium. The patients were discharged after 24 hours post procedure with antiplatelet medication for six months.

Hence, our belief after performing these cases is that transcatheter closure of sinus venosus atrial septal defects can easily be achieved, significantly reducing patient's morbidity as compared to open heart surgery.

Authors Contribution

Muhammad Babar: Conceptualization, data curation, formal analysis, methodology, writing-original draft. Saima Rafique: conceptualization, investigation, methodology. Khurram Akhtar:

conceptualization, resources, writing-review and editing. Maimoona Saeed: Formal analysis. Amjad Mahmood: Formal analysis, validation. Abubakar Sadiq: Methodology, visualization. Azizullah Azizullah: Data curation, investigation, methodology.

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Consent

A written informed consent was taken from each of the patients separately for publication of this case series as well as the accompanying data.

Conflicts of Interest

The authors declare no conflict of interest.

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