

Percutaneous closure of perimembranous ventricular septal defects associated with septal aneurysm in adults

Erişkinlerde septal anevrizmanın eşlik ettiği perimembranöz ventriküler septal defektlerin perkütan kapatılması

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ABSTRACT

Objective: Percutaneous closure of perimembranous ventricular septal defects (pmVSD) has become an accepted alternative to surgical closure in selected cases. However, closure of pmVSDs associated with septal aneurysm is more challenging. We report our experience of device closure of pmVSDs associated with septal aneurysm.

Methods: Between 2008 and 2012, percutaneous closure of pmVSD associated with septal aneurysm was attempted in 11 adult patients in our institution. The patients were followed up at 1, 3, 6, and 12 months after the procedure.

Results: Mean age of the patients (64% male, 36% female) was 36.2±1.3 years. Diameter of the left and right ventricular openings of the aneurysm measured by ventriculography was 13.5±5.6 mm and 5.9±2.2 mm, respectively. The defect was occluded with Amplatzer pmVSD Occluder in 4 patients, Amplatzer Muscular Ventricular Septal Defect Occluder in 4 patients, Amplatzer Duct Occluder I in 1 patient, and Amplatzer Duct Occluder II in 2 patients (AGA Medical Corp., Plymouth, MN, USA). The procedure was successful in all patients. Mean follow-up time was 22±1.9 months. There was no device- or procedure-related complications at the acute setting or mid-term follow-up.

Conclusion: Percutaneous closure of pmVSDs associated with aneurysm is more challenging than that of simple defects. The selection of the device type and size should be made according to the configuration and size of the aneurysm and defect.

ÖZET

Amaç: Perimembranöz ventriküler septal defektlerin (pmVSD) perkütan olarak kapatılması seçilmiş olgularda cerrahi kapatmaya seçenek olarak kabul görmüştür. Ancak septal anevrizmanın eşlik ettiği pmVSD'lerin kapatılması daha zorlayıcıdır. Bu makalede septal anevrizmanın eşlik ettiği pmVSD'lerin cihaz ile kapatılması ile ilgili deneyimlerimizi bildirdik.

Yöntemler: Merkezimizde 2008-2012 yılları arasında septal anevrizmanın eşlik ettiği pmVSD'si olan 11 olguda perkütan kapatma girişiminde bulunuldu. İşlem sonrası 1., 3., 6. ve 12. aylarda hastaların klinik takipleri yapıldı.

Bulgular: Hastaların ortalama yaşı 36.2±1.3 (%64 erkek, %36 kadın) idi. Ventrikülografide interventriküler septumdaki anevrizmanın sol ve sağ ventriküle olan açıklıklarının çapları sırası ile 13.5±5.6 mm ve 5.9±2.2 mm idi. Defektin kapatılmasında 4 hastada Amplatzer pmVSD oklüder cihazı, 4 hastada Amplatzer musküler ventriküler septal defekt oklüder cihazı, 1 hastada Amplatzer Duct oklüder-I cihazı, 2 hastada ise Amplatzer Duct oklüder-II cihazı kullanılması tercih edildi. Bütün hastalarda işlem başarılı olarak tamamlandı. Hastalar işlem sonrası ortalama 22±1.9 ay takip edildi. Erken veya orta dönem takipte cihaz veya işlemlerle ilişkili hiçbir komplikasyon gelişmedi.

Sonuç: Anevrizmanın eşlik ettiği pmVSD'lerde perkütan kapatma basit defektlere göre daha zorlayıcıdır. Cihaz tipinin ve boyutunun seçimi anevrizma ve defektin şekline ve boyutuna göre yapılmalıdır.

Ventricular septal defects are the most common congenital heart defect constituting more than 20% of all congenital heart diseases.^[1] Of these defects, 80% involve the membranous septum. With the

introduction of eccentric perimembranous ventricular septal defect (pmVSD) occluder devices, percutaneous closure of pmVSDs has become an alternative to surgical closure.^[2] However, closure of pmVSDs

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associated with septal aneurysm is more challenging, and various types of devices have been used to occlude these type of defects.^[3-6] We report our experience of device closure of pmVSDs associated with septal aneurysm.

METHODS

Between 2008 and 2012, percutaneous closure of pmVSD associated with septal aneurysm was attempted in 11 adult patients in our institution. The indication for closure was the presence of hemodynamically significant pmVSD demonstrated by cardiac catheterization ($Q_p:Q_s > 1.5$). The defect was examined pre-procedurally by transthoracic (TTE) and transesophageal echocardiography (TEE) to assess the localization and size of the defect and aneurysm, as well as the relationship with respect to the aortic and tricuspid valves. Cardiac catheterization and left ventriculography were performed to calculate the shunt volume and evaluate the defect size and position. The defects causing hemodynamically significant left-to-right shunt that were at least 2 mm away from the aortic valve and localized in the infracristal position were considered suitable for percutaneous closure.

The procedures were performed under general anesthesia with transesophageal echocardiographic and fluoroscopic guidance. The left ventricular opening of the aneurysm was defined as the inlet portion of the aneurysm, and right ventricular opening of the aneurysm was defined as the outlet portion of the aneurysm. The diameter of the left ventricular opening of the aneurysm (the inlet portion) and the diameter of the defect (outlet portion of the aneurysm) were determined by echocardiography and left ventriculography. The closure procedure was performed in accordance with the technique described in previous reports.^[7] The defect was crossed from the left ventricular side by using a right Judkins catheter, and a 0.035" glide wire was advanced to the pulmonary artery. The catheter was then advanced over this wire to the pulmonary artery. The glide wire was exchanged for a 0.035" noodle wire (AGA Medical Corp. Plymouth, MN, USA), which was then snared and exteriorized from the right femoral vein, forming an arteriovenous wire loop. The delivery sheath was advanced over the guide wire from the femoral vein and across the defect into the apex of the left ventricle (LV). The device was loaded and advanced to the left ventricular apex. The distal disc

was deployed in the LV and withdrawn back to the septum. The proximal disc was then opened.

In some cases, it was difficult to position the sheath in the left ventricular apex. In those cases, the sheath was left in the ascending aorta, and the left ventricular disc was opened under the aortic valve. The device was released after confirming the device position and normal function of the aortic and atrioventricular (AV) valves by TEE and angiography.

Four devices were used to occlude pmVSD associated with septal aneurysm: Amplatzer pmVSD Occluder, Amplatzer Muscular Ventricular Septal Defect Occluder Device, Amplatzer Duct Occluder I, and Amplatzer Duct Occluder II (AGA Medical Corp., Plymouth, MN, USA). Patients were prescribed antiplatelet therapy and endocarditis prophylaxis for 6 months.

Patients were followed up at 1, 3, 6, and 12 months after the procedure by TTE and TEE. All patients underwent 24-hour electrocardiography Holter monitoring at 6-month follow-up. The study protocol was approved by the local ethics committee.

RESULTS

Mean age of the patients (64% male, 36% female) was 36.2 ± 1.3 years. Demographic and clinical characteristics of the patients are shown in Table 1. Average diameter of the defect at the outlet portion of the aneurysm was 5.9 ± 2.2 mm (range: 3–9 mm), as measured by angiography. Average diameter of the left ventricular opening of the aneurysm was 13.5 ± 5.6 mm (range: 5–20 mm). One patient had 2 defects within the aneurysm. Amplatzer pmVSD Occluder was used in 4 patients, Amplatzer Muscular Ventricular Septal Defect Occluder in 4 patients, Amplatzer Duct Occluder I in 1 patient, and Amplatzer Duct Occluder II in 2 patients. The closure procedure was successful in all of the patients.

Amplatzer pmVSD Occluder was chosen in 4 patients (Table 1) who had cone-shaped small aneurysms associated with ventricular septal defect. In those cases, there was adequate distance between the

Abbreviations:

AV	Atrioventricular
LV	Left ventricle
PmVSD	Perimembranous ventricular septal defect
RV	Right ventricle
TEE	Transesophageal echocardiography
TTE	Transthoracic echocardiography

Table 1. Demographic, clinical, and procedural characteristics of patients

Patient number	Age (years)	Gender	Aneurysm inlet size (mm)	Defect size (mm)	Distance from the aortic valve (mm)	Qp/Qs	Device
1	26	Male	5	4	2	2.2	PmVSD occluder
2	18	Female	18	8	4	2.0	Muscular VSD occluder
3	36	Male	20	5	8	1.6	Muscular VSD occluder
4	36	Male	19	5	4	1.6	ADO II
5	42	Female	19	8	6	1.7	Muscular VSD occluder
6	62	Male	11	8	8	2.0	PmVSD occluder
7	21	Male	13	9	2	1.7	ADO I
8	49	Female	8	3	7	1.6	PmVSD occluder
9	49	Female	5	3	8	1.7	PmVSD occluder
10	23	Male	14	5	8	2.1	ADO II
11	36	Female	17	8	5	1.7	Muscular VSD occluder

ADO: Amplatzer Duct Occluder; PmVSD: Perimembranous ventricular septal defect.

aortic valve and the left ventricular opening of the aneurysm, with a range of 2–8 mm (Table 1). The defect was closed at the inlet portion of the aneurysm by anchoring the distal disc of the occluder at the left ventricular opening of the aneurysm and aiming to compress the aneurysm and defect between the proximal and distal discs of the device. Some mushrooming appearance towards the right ventricle (RV) was seen in 2 cases (Figure 1).

Amplatzer Muscular Ventricular Septal Defect Occluder was used in 4 patients who had a large circular aneurysm associated with pmVSD. The defect was

closed at the true anatomic hole by anchoring the distal disc of the device at the left side of the outlet portion of the aneurysm, hence placing the device within the aneurysmal pouch (Figure 2).

Amplatzer Duct Occluder I was preferred in 1 patient who had a large tunnel-shaped aneurysm associated with defect, resembling that of a patent ductus arteriosus (Table 1). The distance between the aortic valve and left ventricular opening of the aneurysm was short. The retention skirt of the device was deployed in the ascending aorta and withdrawn back to the septum. The retention skirt was situated within the aneurysm,

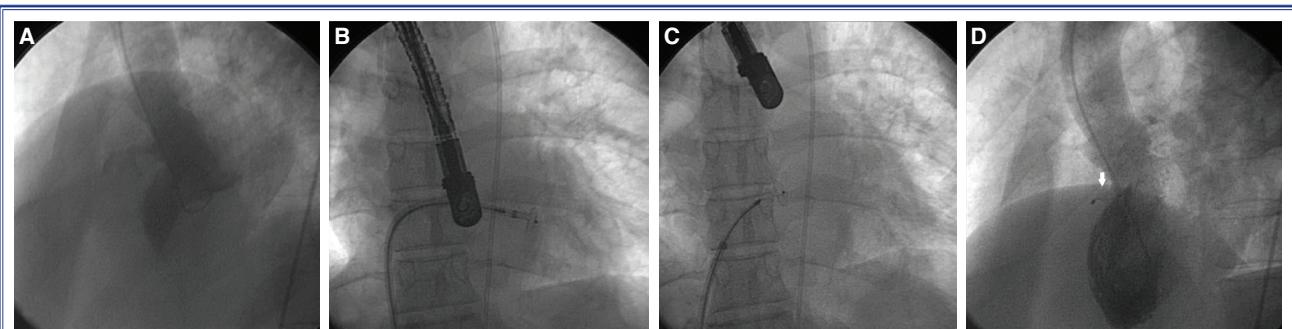


Figure 1. (A) Left ventricular angiogram in the left anterior oblique view, demonstrating pmVSD associated with a small septal aneurysm (Patient 8 in Table 1). **(B)** Cine image during deployment of the left ventricular disc of an 8-mm Amplatzer pmVSD Occluder in the left ventricle. The left ventricular disc is pulled towards the ventricular septum, anchoring the proximal disc at the left ventricular opening of the aneurysm. **(C)** Cine image showing the deployment of the right ventricular disc. **(D)** Left ventricular angiogram after the device has been released, indicating no residual shunt. However, some mushrooming appearance (arrow) of the right ventricular disc has occurred.

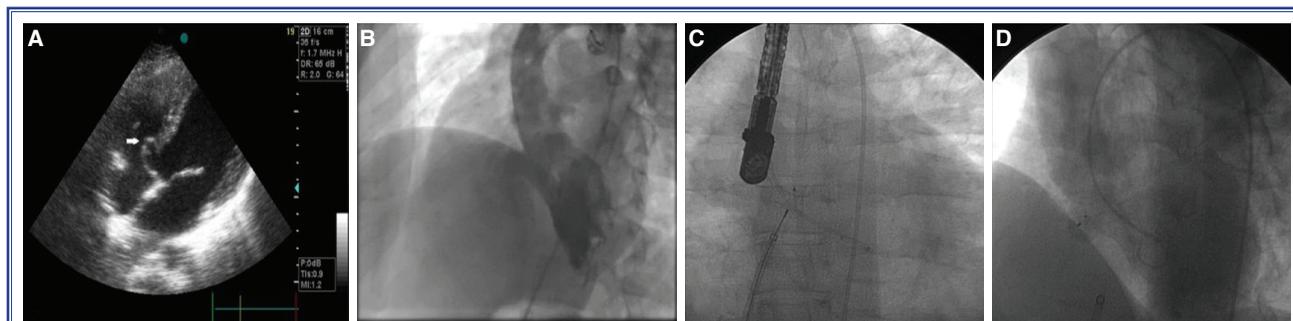


Figure 2. (A) Apical 4-chamber view from transthoracic echocardiography demonstrating a pmVSD (arrow) associated with a large septal aneurysm (Patient 2 in Table 1). (B) Left ventricular angiogram in the left anterior oblique view showing the pmVSD and septal aneurysm. (C) Cine image immediately after deployment of the 10-mm Amplatzer Muscular Ventricular Septal Defect Occluder, occluding the defect at the outlet portion of the aneurysm. (D) Left ventricular angiogram after release of the device, demonstrating that the device is in good position with the left ventricular disc within the aneurysm and there is no residual shunt.

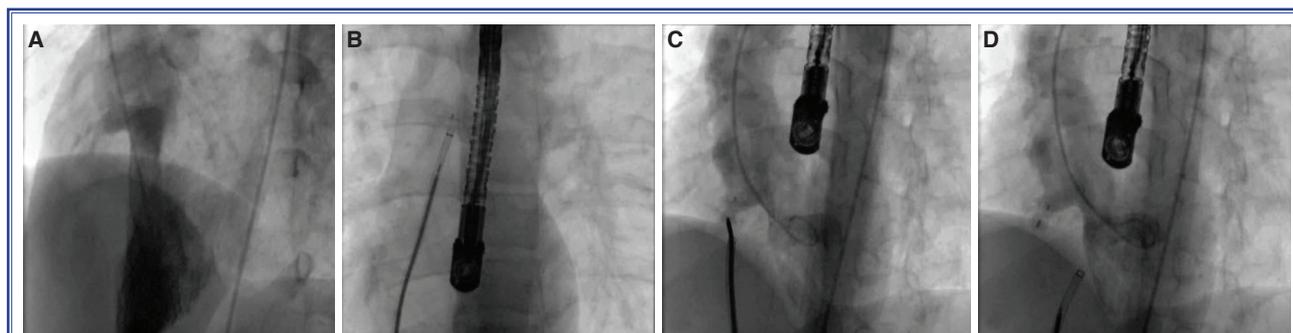


Figure 3. (A) Left ventricular angiogram in the left anterior oblique view demonstrating pmVSD with a tunnel-shaped large septal aneurysm (Patient 7 in Table 1). (B) Cine image showing deployment of the retention disc of the Amplatzer Duct Occluder I in the ascending aorta. The retention disc is then pulled into the aneurysm, and the rest of the device is deployed. (C) Left ventricular angiogram demonstrating the device situated in the septal aneurysm prior to release from delivery cable. (D) Left ventricular angiogram in left anterior oblique view demonstrating the device is in good position and there is no residual shunt.

and the rest of the device was deployed (Figure 3).

Amplatzer Duct Occluder II was used in 2 patients. One of them had dextrocardia associated with pmVSD, which was reported previously,^[8] and the other patient had 2 small defects within the aneurysm. In those cases, a good delivery sheath position in the LV could not be achieved because of the configuration of the defect and aneurysm. Therefore, retrograde approach was used to deploy the device. Using retrograde approach, the distal disc of the device was initially deployed in the RV; accordingly, a device with symmetrical discs was required. Amplatzer Duct Occluder II was preferred in those cases, as it could be delivered through a small delivery catheter.

A trivial residual shunt was detected by ventriculography in 4 patients immediately after implantation of the device. However, complete closure was ob-

served by TTE in all patients at time of discharge. All patients remained in sinus rhythm, and no arrhythmia or conduction defect was observed. There was no dysfunction of the aortic or AV valves. The patients were followed-up at a mean of 22 ± 1.9 months. There was no device- or procedure-related complications at the acute setting or mid-term follow-up.

DISCUSSION

Perimembranous ventricular septal defect is the most common congenital heart disease, and percutaneous closure has become an alternative to surgical closure in most cases. However, the proximity of the margins of the pmVSD to the conduction system, aortic valve, and AV valves increases the risk of adverse events related to percutaneous closure.^[2] The presence of aneurysmal tissue around a pmVSD makes the morphol-

ogy and device selection even more complex. Several types of devices and techniques have been used for percutaneous closure of pmVSDs associated with septal aneurysm. Occlusion of these type of defects with the eccentric pmVSD occluder have been attempted, but disfigurement of the device has been reported.^[3,9] Mushrooming appearance of the body and right disc can occur when a pmVSD occluder is used in patients with septal aneurysm. Similar to those reported cases, mushrooming effect was observed in 2 patients in the present study whose defect was occluded with a pmVSD occluder at the left ventricular opening of the aneurysm.

In cases with a large aneurysmal pouch which seems to accommodate the left ventricular disc of the occluder device, the defect was closed at the outlet portion of the aneurysm by anchoring the left ventricular disc of the occluder at the left side of the outlet portion of the aneurysm. The type of occluder device was selected based on the size and shape of the aneurysm and the defect. The Amplatzer Muscular Ventricular Septal Defect Occluder was used in patients who had a large circular aneurysm. Diameter of the aneurysmal pouch was taken into consideration to be certain that the aneurysm would accommodate the distal disc of the muscular ventricular septal defect occluder device.

The morphology of some pmVSDs associated with aneurysm resembles that of patent ductus arteriosus, with a large opening on the left ventricular side and a restrictive opening on the right ventricular side. Therefore, occlusion of these types of defects with devices designed for patent ductus arteriosus has been attempted in a small number of patients.^[4,6,10] We chose to use the Amplatzer Duct Occluder I in a patient with a tunnel-shaped aneurysm. The retention skirt was implanted within the aneurysm, and the cylindrical portion was secured in the opening of the aneurysm to the RV. El Said et al. reported that suitability of the Amplatzer Duct Occluder I was assessed according to the configuration of the aneurysm and defect.^[6] Amplatzer Duct Occluder I was found to be suitable if the orientation of the right ventricular opening of the aneurysm was at an acute angle to the interventricular septum. Although long-term follow-up data is not available for the Amplatzer Duct Occluder I, no device disfigurement or conduction block was reported in the acute setting or at mid-term follow-up.^[6]

Amplatzer Duct Occluder II was used in a patient with multiple holes within the aneurysm and in a patient with dextrocardia. Because of the configuration of the defect and aneurysm, retrograde approach was used to deploy the device. Therefore, Amplatzer Duct Occluder II with symmetrical discs was preferred. Having symmetrical discs, Amplatzer Muscular Ventricular Septal Defect Occluder might have been an alternative in those cases. However, Amplatzer Duct Occluder II can be delivered through a smaller delivery sheath, which could be advantageous in manipulating the delivery system and the device.

Implantation of the occluder device at the right ventricular opening of the aneurysm places the device within the aneurysm at a distance remote from the aortic valve and conduction tissue. This may be related to decreased risk of AV block and interference with the aortic valve. Accordingly, the absence of conduction disorders in our patients as well as those reported in previous studies^[5,6] might be related to the implantation of the device away from the conduction tissue.

Limitations

This study is limited by its small sample size. Larger series with longer follow-up data are needed to reach conclusions regarding the suitability, efficacy, and safety of each device.

In conclusion, percutaneous closure of pmVSDs associated with septal aneurysm is feasible, but pmVSD occluder devices seem to be unsuitable for these defects. However, if the aneurysm is small, the device could cover the hole and aneurysmal tissue. In cases of large aneurysms, implantation of the device within the aneurysm and occlusion of the true anatomic hole might be considered. Selection of the device should be made according to the configuration and size of the aneurysm and anatomic hole.

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Keywords: Congenital heart disease; percutaneous closure; ventricular septal defect.

Anahtar sözcükler: Konjenital kalp hastalığı; perkütan kapatma; ventriküler septal defekt.