Transcatheter device closure of perimembranous ventricular septal defect associated with indirect Gerbode defect: A retrospective study

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ABSTRACT

Perimembranous ventricular septal defect (pmVSD) is a common congenital heart disease that is sometimes associated with indirect left ventricle (LV) to right atrium (RA) shunt (indirect Gerbode defect). This defect has a rare chance of spontaneous closure and therefore was usually closed surgically in the past, but more recently transcatheter closure has been reported by a few authors. In our study, we have described a series of 14 children (age ranging from 1.2 to 12 years and weight ranging from 7.2 to 25.5 kg) with the above-mentioned defect which were closed by various interventional devices. The procedures were successful in complete elimination of pmVSD and immediate reduction of indirect LV-RA shunts with negligible residual tricuspid regurgitation on follow-up. In our midterm experience, the judicious use of double-disc devices is efficacious for occluding pmVSD associated with indirect Gerbode defect.

Keywords: Amplatzer duct occluder II device, gerbode defect, percutaneous device closure, perimembranous ventricular septal defect

INTRODUCTION

Ventricular septal defect (VSD) is the most common type of congenital heart disease.[1] Of these, perimembranous VSD (pmVSD) forms the most common subtype, which is occasionally associated with indirect left ventricle (LV) – right atrium (RA) shunt. This phenomenon has also been referred to as indirect type of Gerbode defect.[2,3] This study aims at a retrospective analysis of feasibility and midterm outcome of transcatheter closure of this lesion using double-disc devices in children.

MATERIALS AND METHODS

In this retrospective study, among all the VSD device closures performed on children between November 2017 and March 2020, we included patients who had a significant LV-RA shunt through a pmVSD for analysis. Most of our cases initially presented with pmVSD and high-velocity tricuspid regurgitation (TR) but careful transthoracic echocardiography (TTE) showed that there was a well-developed aneurysm formed by septal tricuspid leaflet (STL) covering the actual defect, with or without adherence of anterior tricuspid leaflet (ATL) to the VSD margin. This associated defect in the tricuspid valve apparatus resulted in partial shunt of the VSD jet into RA which appeared as TR [Figure 1]. The morphology and effective size of VSD, the severity of indirect LV-RA shunt, any chamber dilation, and presence of pulmonary artery (PA) hypertension were thoroughly scrutinized. The effective size of VSD and exit points were determined by 2D and color Doppler

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echocardiography as the narrowest diameter of orifice in the aneurysmal VSD tract and an average of three such measurements was recorded. The severity of this indirect LV-RA shunt appearing as TR was classified into mild, moderate, or severe according to the American Society of Echocardiography recommendations.[4] Furthermore, the chest X-ray and electrocardiogram (ECG) reports of the patients were evaluated for cardiomegaly and signs of LV volume overload, respectively.

Following the above-mentioned criteria, we selected a total of 14 cases and carefully analyzed their records, catheterization report, and follow-up data. The catheterization procedure was performed under local anesthesia and intravenous sedation. The LV angiogram delineated the VSD jet and indirect LV-to-RA shunt in most cases [Figure 2]. The VSD was crossed retrogradely with the help of an exchange length wire and PA was entered with a diagnostic catheter to record the PA pressure. Appropriate-sized devices (KONAR-MF VSD Occluder in one case and Amplatzer Duct Occluder [ADO] II in the rest) were deployed through a guide catheter under echocardiographic and angiographic guidance.

However, in one case, the effective VSD size was borderline and even the largest available ADO II device slipped off through the defect upon deployment. In this case, therefore, an arteriovenous loop was formed across the defect and we deployed an ADO I device in an antegrade manner. In all the cases, a detailed TTE was done before releasing the device to ensure its position across the defect and that there is no significant residual shunt.

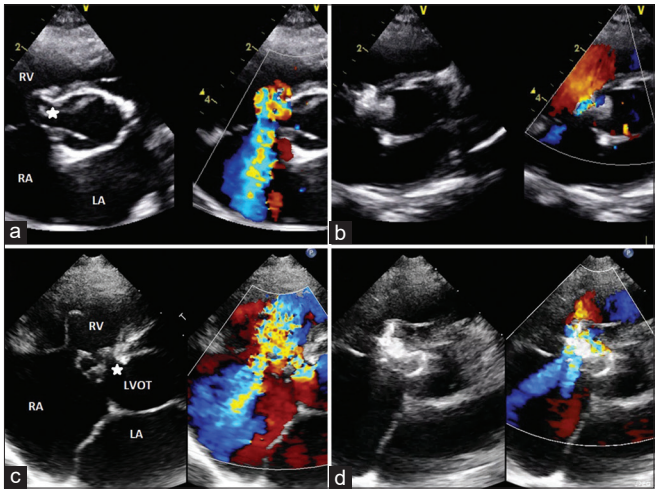

Figure 1: Echocardiographic pictures (pre and postintervention). One case with baseline severe indirect Gerbode shunt (a) which showed no residual VSD and trivial tricuspid regurgitation immediately after device closure (b) Another case with well-developed tricuspid septal leaflet aneurysm (depicted by asterisk) and severe indirect Gerbode shunt (c) showing small residual VSD and moderate tricuspid regurgitation after the procedure (d). LA: Left atrium; RA: Right atrium; RV: Right ventricle; LVOT: Left ventricle outflow tract

Postprocedure the patients were discharged after documenting a satisfactory 12 lead ECG and TTE examination. On discharge, we didn't prescribe aspirin according to our hospital policy but bacterial endocarditis prophylaxis was recommended for the next 6 months. Further follow-up was planned for echocardiography and ECG after one, 6, and 12 months and then annually afterward.

RESULTS

There were six male and eight female patients included in this study and their baseline characteristics are depicted in Table 1. The median age at the time of intervention was 8 years (range 1.2–12 years) and the mean weight was 17.4 kg (range 7.2–25.5 kg). The median duration of follow-up was 16.5 months. Cardiomegaly in chest X-ray (cardiothoracic ratio ≥ 0.6) was noted in 6 cases, while LV volume overload by echocardiography (LV end-diastolic diameter ≥+2.5 standard deviation) was present in 9 out of 14 cases. The mean fluoroscopy time during the procedure was 9.9 min. The mean PA pressure was 23.5 mmHg (range from 18 to 27 mmHg). The mean size of an effective VSD orifice was 4.6 mm (range from 3.5 to 6.5 mm). The preprocedural severity of indirect LV-RA shunt was severe in five, moderate in seven, and mild in the remaining two cases.

Device placement was successful in all 14 cases with no immediate procedural complications. We classified residual VSD according to its jet width in color Doppler TTE into trivial (<1 mm), small (1–2 mm), moderate (2–4 mm), or large (>4 mm). According to these criteria, we documented trivial and small residual

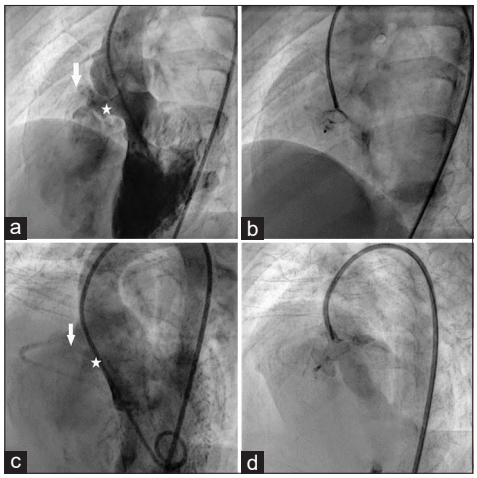

Figure 2: Angiographic pictures before and after the device deployment. One case with pmVSD (depicted by asterisk) restricted with well-developed septal aneurysm and faintly visible indirect left ventricle-to-right atrium shunt (arrow) (a) which was closed effectively with an AMPLATZER™ Duct Occluder II device (b) Another similar case (c) being intervened successfully with a KONAR-MF™ VSD Occluder device (d)

Table 1: Patient characteristics, device type and size, and outcome of procedure

Age (years) at procedure/ sex	Weight (kg) at procedure	Indications for intervention	Effective VSD size (mm)	Baseline severity of TR	Device type and size (mm)	PP* residual VSD	PP* severity of TR	Follow-up severity of TR†	Follow-up duration (month)
1.2/female	7.2	FTT, recurrent LRTI	5	Moderate	MF VSDO (8/6)‡	Trivial	Mild	Mild	1
4/female	14.3	LA and LV dilation	3.5	Mild	ADO II (4/4)§	None	None	None	11
4.5/male	12.7	FTT, exercise intolerance	6	Severe	ADO II (6/6)	Trivial	Mild	None	11
5/male	16.2	LA and LV dilation	4.5	Moderate	ADO II (6/4)	None	Mild	None	21
5/male	14.7	FTT	4	Moderate	ADO II (5/4)	None	None	None	15
6/female	15.3	LA and LV dilation	4	Moderate	ADO II (5/4)	None	Mild	None	23
8/male	19.0	Exercise intolerance	4	Moderate	ADO II (5/6)	None	Mild	None	18
8/male	19.5	LA and LV dilation	4	Moderate	ADO II (5/4)	None	Mild	None	14
8.5/female	21	Exercise intolerance	6.5	Severe	ADO I (10/8)‖	Small	Moderate	Moderate	21
9/female	14.7	FTT, recurrent LRTI	5	Severe	ADO II (6/6)	Trivial	None	None	20
9/female	18.4	Exercise intolerance	4	Severe	ADO II (5/4)	Trivial	Mild	None	11
9/male	22	LA and LV dilation	4	Mild	ADO II (5/4)	None	None	None	10
11/female	23.4	Exercise intolerance	4.5	moderate	ADO II (5/6)	None	Mild	None	19
12/female	25.5	Exercise intolerance	5.5	Severe	ADO II (6/6)	Small	Moderate	Mild	22

*Immediate postprocedure value, †The last follow-up of the patient after the procedure, ‡MF VSDO (x/y): KONAR-MF™ VSD Occluder; x=waist diameter at LV side (mm) and y=waist diameter at RV side (mm), §ADO II (x/y): ADO II; x=waist diameter (mm) and y=nominal length (mm), ||ADO (x/y): ADO; x=device diameter at descending aorta (mm) and y=device diameter at pulmonary artery (mm). FTT: Failure to thrive, LRTI: Lower respiratory tract infection, LA: Left atrium, LV: Left ventricle, TR: Tricuspid regurgitation due to indirect Gerbode defect, Years: Years of age, VSD: Ventricular septal defect, ADO II: AMPLATZER™ Duct Occluder II, VSDO: Ventricular septal defect occluder

VSD in four and two cases respectively immediately after the procedure [Figure 1], which disappeared completely on 6 months follow-up. All cases showed an immediate reduction in the severity of TR to mild or less except in one case where we had used the ADO I device. In this case, although moderate TR persisted even after 1 year of follow-up, the patient remained asymptomatic on low dose diuretic medication. None of the patients developed any complications like atrioventricular conduction block, hemolysis, thromboembolic event, or device embolism. However, two patients developed loss of pulse in the limb used for procedural access which resolved with heparin infusion and all the patients were discharged satisfactorily.

DISCUSSION

There are several studies that have meticulously examined into the mechanism of aneurysmal transformation of STL with or without any involvement of ATL, and their contribution in LV-to-RA shunt.[5,6] According to one study, the majority of untreated cases with pmVSD would eventually develop aneurysmal transformation, and about 11% of these will have indirect LV-to-RA shunt.[6] The additional mechanism of this shunt appears to be related to the VSD jet pushing the ATL away from its coaptation surface with STL.[7] Spontaneous closure of this defect is rare, and it is often followed by worsening of the TR unless early surgery or intervention is done.[6] Surgical correction was the method of choice till recently, when a few isolated reports of transcatheter closure of both direct and indirect types of Gerbode defect emerged.[8-10] Another study by Kerst *et al.* comprising of four children with pmVSD and indirect LV-RA shunt elucidated their successful transcatheter closure.[11] However, the most comprehensive case series was published by Vijayalakshmi *et al.* where intervention on 12 patients with Gerbode defect, half of which were of indirect type, was done successfully with ADO II devices though with some incidence of heart block.[12]

In our experience, double-disc low profile devices are well suited for the closure of indirect Gerbode defect associated with pmVSD in terms of significant reduction or complete elimination of LV-RV and indirect LV-RA shunts. In most of our cases, the landing zone of the device was deep-seated in the aneurysmal VSD tunnel, and well away from the conduction system. Therefore, we did not experience any incidence of heart block or aortic valve impingement by the devices used. With proper case selection, there should be minimal risk of immediate and long-term complications in hands of experienced operators.

Limitations

The number of patients in our study as well as the size and morphology of the defect is inadequate for any generalized conclusion. We need more evidence with long-term follow-up data of similar procedures.

CONCLUSIONS

The indirect Gerbode defect associated with pmVSD should not be a contraindication for device closure if the case is otherwise suitable for the procedure. Double disc devices, like ADO II, appear promising for the correction of this defect.

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Conflicts of interest

There are no conflicts of interest.

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