

Cardiac rhythm abnormalities after transcatheter device closure of perimembraneous ventricular septal defects in pediatric patients at intermediate term follow up

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Abstract

Background: Transcatheter closure of perimembraneous ventricular septal defects in pediatric patients poses special challenges owing to anatomical relationship between conduction system and perimembraneous ventricular septal defect. **Aims:** The study aims to assess and evaluate the conduction disturbances after transcatheter device closure of perimembraneous ventricular septal defect using different occluders at mid-term follow-up. **Methods:** We studied 297 patients having PMVSD with clinical and/or echocardiographic evidence of a significant left-to-right shunt. All subjects underwent clinical examination, electrocardiogram (ECG), chest X-rays and transthoracic echocardiography before discharge and at 1, 6 and 12-months after the procedure and yearly thereafter. Platelet anti-aggregation therapy with aspirin 5 mg/kg/day orally and endocarditis prophylaxis was prescribed for six months. **Results:** The mean age of the patients was 8.64±3.14 years (range 3-17.2 years). Majority (86.2%) had no residual shunt at follow-up. Total rhythm disturbances were seen in about 6% (18/297) of patients with transient complete atrioventricular block (CAVB) occurring in 3 patients. There was no mortality in our study which compares well with the surgical results in which it is between 0-3 percent. **Conclusions:** This study showed that transcatheter closure of PMVSD using symmetric PMVSD occluders and duct occluders is a safe and effective alternative to surgery. Oversizing of devices should be minimized and low profile devices appropriate to specific morphology of VSD should be used. It has excellent results in experienced hands with minimum morbidity and almost no mortality.

Keywords: Structural heart disease, Perimembraneous ventricular septal defect, Transcatheter closure, Pediatric cardiology

Introduction

Isolated ventricular septal defect (VSD) is the third most common congenital cardiac malformation after bicuspid aortic valve and mitral valve prolapse. VSDs account for 20 – 30% of the congenital heart defects. The most common type is the perimembraneous PMVSD accounting for 70% of all VSDs [1]. PM VSDs are those opening in the region of the inner curvature of the right ventricle, having as their diagnostic feature the continuity between the leaflets of tricuspid and aortic valves, which also incorporates the central fibrous body [1, 2]. Indications of VSD closure are symptoms of heart failure, signs of left ventricular

volume overload, and history of endocarditis. Standard treatment for PM VSD is open surgery, which is widely performed with minimal operative mortality but still carries risks, such as residual shunt, post-pericardiotomy syndrome, wound infection, scar etc. In the last two decades, percutaneous techniques to close cardiac defects have been developed [3, 4]. Closure of atrial septal defects, patent ductus arteriosus, and muscular ventricular septal defects using transcatheter devices has been widely reported. Owing to anatomical relationship of conduction system and PMVSD location, and reported high incidence of conduction disturbances after device closure, it is prudent to ensure clinical and electrocardiographic follow up for prolonged duration [5]. While, recently published

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studies have shown encouraging results of PMVSD device closure, long-term follow-up results are still awaited [7, 8]. This study aims to evaluate the safety and efficacy of transcatheter device closure of PMVSD with special emphasis on conduction disturbances.

Materials and Methods

This is a single centre prospective study which included 297 pediatric patients between 2010 and 2014 who underwent percutaneous PMVSD device closure at our institution. All patients were screened by conventional two dimensional and color doppler transthoracic echocardiography. Before intervention an informed written consent was obtained from all patients/or parents.

Detailed history was taken including any previous history of infective endocarditis. Other associated non cardiac anomalies were also ruled out. General physical examination, standard 12- lead ECG, chest x-ray and routine blood investigations including complete blood counts, serum electrolytes, renal function tests and HIV and Hep B serologies were done. 24 hr Holter monitoring was done using SEER Light digital recorder (7 lead, 3 channel) GE Healthcare systems, at a minimum follow up period of 6 months or earlier if required. In addition to conduction disturbances any associated symptoms or tachyarrhythmias if present, were also noted during the period of Holter monitoring.

Results

Patient-related variables- Transcatheter closure of PMVSD was attempted in 297 patients which was successful in 295 patients (99%). There were 170 male and 127 female patients. The age ranged from 3 years to 17.2 years (8.64 ± 3.14). The weight varied between 10 kg upto 63 kg (21.11 ± 8.26). The defect size on two dimensional echocardiography varied between 2.5mm to 10.2 mm (4.98 ± 1.41).

The diameter of the VSD was measured by transthoracic echocardiography using 2-D imaging and color flow Doppler on long-and short-axis views. The vertical diameter was measured during left ventricular angiography in LAO cranial views. In case of ventricular septal aneurysm VSD size was also measured on the left ventricular side. Catheterization procedure was performed under local or general anaesthesia. Arterial and venous access were taken. Heparin 100U/kg and antibiotics were administered intravenously before procedure. All subjects underwent clinical examination, ECG, chest X-rays and TTE before discharge and at 1, 6 and 12 months after the procedure and yearly thereafter. Platelet anti-aggregation therapy with aspirin 5mg/kg/day orally and endocarditis prophylaxis was prescribed for 6 months. Holter monitoring was done at a lag period of 6 months or earlier if required. Oral steroids (tab prednisolone 1-2mg/kg) were given to all patients and dose was tapered over 2-3 week period. Treatment with steroid was extended in patients developing post procedure conduction disturbances if required.

Statistical Analysis- Data were expressed as a frequency or percentage for nominal variables, as the median (range) for categorical variables and as mean \pm SD or median (range) for continuous variables. A paired t test was performed as indicated to compare two mean values. $P < 0.05$ was set as the level of statistical significance.

Table-1

Parameters	Mean	Standard deviation	Range
Age (years)	8.643	3.1433	3.0-17.2
Weight (kg)	21.11	8.269	8-63
Height (cm)	125.11	17.414	84-174
BSA (m ²)	0.85	0.22	0.47-1.74
Device size (mm)	7.79	2.08	4-16
VSD size (mm)	4.98	1.42	2.5-10.20
Device/Defect ratio	1.35	0.35	1.1-2.9
Follow up (months)	13.39	12.67	1-63
Fluoroscopy time(min)	12.98	8.649	3-70
Delivery system (Fr size)	7.79	2.08	4-16

Demographic and catheterization data of pediatric patients who underwent VSD device closure

Catheterization data- Cardi-O-Fix PM VSD occluder (Starway Medical Technology Inc., Beijing) was the most commonly used device (60% patients) followed by Shanghai Symmetrical PM VSD occluder (Shape Memory Alloy Ltd, Shanghai, China) in 30% patients, Cardi-O-Fix ductal occluder in 9% patients and Amplatzer ductal occluder (St. Jude Medical Inc., Minnesota, USA) in 1 % patients. The size of device used varied between 4 to 16mm (7.79 ± 2.08). The fluoroscopy time of procedure ranged between 3min to 70 min (12.98 ± 8.64) while mean device/defect ratio varied between 1.1 to 2.9 (1.35 ± 0.35).

Complications- There were only two major complications including one case of device embolization to right pulmonary artery. The patient was asymptomatic with no evidence of vascular compromise of right lung. The VSD was closed electively by surgery later. The second patient had persistent gross hematuria following device closure of VSD which was managed with blood transfusion and supportive treatment. Subsequently device retrieval was done followed by surgical closure. All the other complications were minor including transient loss of pulses, local site hematoma, fever etc. There was no correlation of complications with the weight, age or size of the device or delivery system used in the patients.

Cardiac rhythm disturbances were seen in about 6% (18/297) of patients with transient CHB occurring in 3 patients. Conduction disturbances were transient in most of the patients. Overall, most common conduction disturbances encountered in decreasing order of frequency were junctional rhythm (6 patients), RBBB complete and incomplete (3 patients each), incomplete LBBB (1 patient), complete LBBB with first degree AV block (1 patient) and bifascicular block in 1 patient.

Discussion

Perimembranous VSDs are one of the most common congenital heart defects [1-3]. Patients with volume overload of the left ventricle due to a VSD require closure of the defect to prevent ventricular dilatation and dysfunction, arrhythmias, aortic regurgitation, pulmonary arterial hypertension, endocarditis and double chambered right ventricle [4]. Standard treatment for PMVSD is surgery which is considered a safe procedure but has some potential complications like residual shunts [5-8], necessity of reoperation in 2 % patients and even death in 0.6 – 5% cases [5,7-10]. The incidence of complete heart block after surgery has been around 3% in earlier studies which has fallen further with advancement in techniques [11].

Percutaneous approach for the treatment of congenital heart diseases is favoured by patients and their parents because it has less psychological impact (given the absence of a skin scar), and hospital stay is shorter. The procedure also causes less pain and discomfort and there is no need for admission to an intensive care unit. This is one of largest single centre prospective studies to evaluate the intermediate term outcomes of percutaneous PM VSD device closure in pediatric patients with success rate of 99%. This is consistent with other studies where success rate ranged between 87-100% [12-16]. There was no mortality in our study which compares well with the surgical results in which it is between 0-3% [17-20]. In our study, the total complication rate was 19.5% (most of them

minor) with majority of the patients having transient loss of peripheral pulses, hematoma etc. However, major complications occurred in only <1% of the cases which is consistent with data from other studies worldwide [15-19] in the multivariate analysis factors associated with the occurrence of complications were size of delivery system and fluoroscopy time.

No significant aortic regurgitation occurred in our series even though we used symmetric PM VSD occluders and duct occluders which have disc of 2 mm towards the aortic side. The explanation could be the exclusion of patients with aortic rim <4 mm in our study. The incidence of residual shunting, which was 13.8% at discharge, decreased to 1.3% during follow-up. Residual shunting was not associated with any type of specific occluder used. In all the subjects, the residual shunt was either trivial or mild. TR was moderate to severe in 6.4% cases and was strongly associated with STL closure of the VSD. The incidence of wasting (PEM - all grades) was 85 % in our study with stunting present in only 11.4% of the patients. PAH was present in two third of the patients and strongly correlated with Qp/Qs ratio of >2.

In patients treated percutaneously, the occurrence of conduction disturbances and CHB is quite unpredictable. This complication is related to the proximity of the conduction system to the margins of

the PMVSD. Therefore, both surgery and device implantation may interfere with atrioventricular conduction. Various mechanisms may be considered as causative. It is possible that the presence of the device may disturb atrioventricular conduction by direct traumatic compression. Furthermore, the device may give rise to an inflammatory reaction or scar formation in the conduction tissue.

However, there is no specific data about the mechanisms involved in the occurrence of CHB after percutaneous closure of a PMVSD. Initially with introduction Amplatzer muscular VSD occluder, there was high incidence of CHB despite high rates of successful VSD closure. With its small waist and small aortic disc (0.5mm) Amplatzer device was specifically designed for closure of PM VSD and could be used for defects with aortic rims as small as 2mm. But the LV disc was larger (5mm) so as to provide stability to the device. With its large LV disc and small waist there had been more compression of the septum and thus the conduction system. Secondly, most of the operators were in the initial learning phase.

VSD with inlet extension and Down's syndrome were other factors associated with CHB. Nearly, 20% of these patients had late onset CHB. Many of these patients had early inappropriate permanent pacemaker implantation mainly due to lacunae in the knowledge of natural history of CHB in these patients. In these studies two third of the patients were either less than 2 years old or had weight < 8 kg. Studies showed patient age and weight to be an independent risk factor of CHB [21].

The myocardium in patients <3 years old is more immature, characterized by increased water content and a less compact structure, which predisposes to myocardial edema leading to CHB. The success rate in our study in our study can be attributed to careful exclusion of patients less than 3 years of age in whom surgical repair may be the preferred option.

The low incidence of conduction disturbances with symmetrical VSD occluders can be attributed primarily to smaller LV disc and relatively longer waist as compared to Amplatzer VSD occluders. The incidence of conduction disturbances was 6.1% in our study which is much lower than reported in the previous studies [6-8]. Most of the conduction disturbances were transient with recovery occurring within one week of the procedure. Complete atrioventricular block occurred

in three patients in our study but none of them required permanent pacing. There was a specific pattern of conduction disturbances seen in most of the patients particularly those with junctional rhythm. Almost all the patients with junctional rhythm developed this abnormality between 1st and 3rd post procedure days with return of normal conduction by 7th post procedure day. Rhythm disturbances were permanent in 4 patients including incomplete RBBB in 2 patients and incomplete LBBB and complete RBBB in 1 patient each.

There was no significant correlation between the size of device or delivery system and the conduction disturbances. Most of the patients in our study with conduction disturbances responded to steroids with recovery occurring in most of the patients. Steroids help in ameliorating the edema and inflammation due to impingement of device and delivery system on the septum and thus the conduction system.

The lower mean fluoroscopy time and lower incidence of repeat maneuvers in our study can also account for the low incidence of conduction abnormalities.

Conclusions

Our study shows that transcatheter closure of perimembraneous VSD using symmetric PMVSD occluders and duct occluders is a safe and effective alternative to surgery. It has excellent results in experienced hands with minimum morbidity and almost no mortality. But onset of atrioventricular conduction disturbances are unpredictable and patients should be followed up carefully till the post procedure ECG returns to normal. Oversizing of devices should be minimized and low profile devices appropriate to specific morphology of VSD should be used. Adverse events including serious conduction disturbances are rare but also manageable.

The use of oral steroids over a short term period can drastically reduce the incidence of conduction disturbances. The decision to implant a permanent pacemaker in the absence of guidelines should only be done only after optimized anti-inflammatory regimen for 3-4 weeks. Similarly, incidence of post implant complications can be minimized by excluding small patients and selecting appropriate kind of occlusive devices. Newer hardware design or modifications like steroid eluting devices may reduce the incidence of conduction disturbances.

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Abbreviations

PMVSD – perimembraneous ventricular septal defect, CHB - complete heart block, LAO - Left anterior oblique, mLBBB - Left bundle branch block, RBBB - Right bundle branch block

Conflict of Interest: The authors declare that they have no conflict of interest. Informed consent was obtained from all individual participants included in the study.

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