DOUBLY COMMITTED SUB ARTERIAL VENTRICULAR SEPTAL DEFECT DEVICE CLOSURE USING NIT-OCCLUD® PDA-R DEVICE: INITIAL EXPERIENCE IN PAKISTAN

Amjad Mehmood, Khurram Akhter, Nadeem Sadiq, Mehboob Sultan*, Hajira Akbar

Armed Forces Institute of Cardiology/National Institute of Heart Disease (AFIC/NIHD)/National University of Medical Sciences (NUMS) Rawalpindi Pakistan, *Army Cardiac Centre, Lahore Pakistan

ABSTRACT

Objective: To assess the suitability of transcatheter closure of doubly committed subarterial ventricular septal defect (DCSAVSD) with PDA-R device for the first time in Pakistan.

Study Design: Descriptive cross-sectional study.

Place and Duration of Study: Paeds Cardiology department AFIC/NIHD Rawalpindi, from Dec 2018 to Jul 2019.

Methodology: A total of ten patients with DCSAVSD more than three years of age with no other associated congenital cardiac malformations were subjected to device closure. Few patients had mild aortic valve regurgitation. Device used was PDA-R pfm-nitocclud which is preloaded to the delivery system. The procedure was carried out under general anesthesia and monitoring of heart rate and hemodynamics. Preliminary detailed echocardiogram was done to assess the suitability of device closure. All patients were followed post procedure.

Results: Complete closure was attained in all patients with no residual flow. No mortality was faced. Two patients had brief episode of heat block during the arteriovenous loop creation which subsided at the end of procedure.

Conclusion: Patients with DCSAVSD can be closed effectively with PDA-R device. Selection of cases should be adequate matching the VSD with the device size. This procedure is good alternate to surgical closure avoiding scar, cost and hospital stay.

Keywords: Aortic regurgitation, Doubly committed subarterial ventricular septal defect, PDA-R.

INTROUCTION

Doubly committed subarterial ventricular septal defect (DCSAVSD) comprises about 30% among all ventricular septal defects (VSDs) types especially in Asian population in comparison with western world (10%)[1]. This is a unique type of defect where spontaneous closure is very uncommon. However DCSAVSD favors coronary cusp prolapse due to venture effect which leads to aortic valve incompetence. This feature necessitates closure of DCSAVSD[2]. The infundibular (or conus) septum separates the tricuspid and pulmonary valves and accounts for the more superior placement of the pulmonary valve relative to the aortic valve. This portion of the septum also provides fairly rigid, muscular support for the aortic valve, especially the right coronary cusp[3]. Extracardiac mesenchyme, derived from neural crest tissue, condenses as prongs (which act as a welding agent) with the most superior portion of the distal cushions to form the aorto-pulmonary septum[4]. The frequent association between arch abnormalities and significant conal VSDs suggests a common mechanism involving a chromosome band 22q11 microdeletion. Deletions in this area have not been linked with isolated supracristal VSD[5].

Device closure of DCSAVSD is attempted for the first time at our center (Armed forces institute of cardiology, Rawalpindi, Pakistan). Although this procedure has been carried out at various centres worldwide but our experience of closing the defect transcatheter has shown remarkable results with 100% success rate so far.

There is no specific device designed for occlusion of DCSAVSD but we have used three different kinds of devices for this purpose[14]. The

Correspondence: Dr Amjad Mehmood, Department of Cardiology, AFIC/NIHD Rawalpindi Pakistan
Email: amjpaedcard@yahoo.com

main concern is entrapment of right ventricular (RV) tissues, blockage of right ventricular outflow tract (RVOT) while implanting RV end of the device and peri procedure injury to conduction pathway causing arrhythmias[15]. To avoid these complications we used Nit-Occlud® PDA-R devices (pfm medical) keeping in view the harmless RV end during and after implantation and very soft consistency of the device which is very gentle during maneuvering across the aortic valve leaflets.

METHODOLOGY

From December 2018 to July 2019, ten patients of DCSAVSD were selected for device closure. Among these 8 were males and 2 were females. DCSAVSD with other congenital cardiac lesions requiring surgery were excluded for device closure. Necessary approval was sought from the institute. All patients were thoroughly screened by echocardiogram assessing the size and exact location of defect, color flow and continued wave Doppler across VSD, relation to pulmonary artery and RVOT tissue, aortic valve leaflets morphology, presence and extent of leaflet prolapse, presence and severity of aortic valve competence, tricuspid and pulmonary valve morphology, Pulmonary artery pressures and other associated lesions. Very large defects with severe pulmonary hypertension and less than three years of age were not considered suitable for transcatheter device closure.

Out of 10 patients 4 had right coronary cusp (RCC) prolapse and only one patient was observed having mild aortic regurgitation. The selected nit-occlud® PDA-R pfm medical device (fig-1) was presumed to close the defect as well as approximate the RCC and minimize valve incompetence.

During preliminary echocardiogram the size of the defect dictated the size of the device which was estimated to be double of the narrowest point of the defect. For proper placement and closure of the defect all size PDA-R devices were made available. In order to have minimum maneuvering of the device during disengagement at the end of procedure the cable at the distal end of delivery system is gently pulled straight back. Left ventricular (LV) angiograms were done to properly asses the placement of the device, any residual leakage across the defect, proximity of the device to the defect and presence of aortic regurgitation. All selected patients were subjected to preliminary investigations for anesthesia fitness. Anemia, infection including hepatitis screening and coagulopathies were ruled out prior to procedure. Any patient not fit for general anesthesia was postponed till normality was ascertained.

Procedure of Device Closure

The procedure of transcatheter device closure of DCSAVSD was done under general anesthesia using laryngeal mask. Femoral artery and venous sheaths were introduced and femoral artery was flushed with heparin 50 units/Kg to avoid thrombosis. LV angiogram was done in three standard projections left anterior oblique (LAO) 25 and cranial 25 degrees, LAO 90 degree, RAO 30 degree to clearly define the size, site, extent and relation to aortic valve. VSD is crossed from LV side of the defect most easily in LAO 90 projection with extra length terumo wire. Although sometimes it's difficult to cross the defect and gentle maneuvering of terumo wire is recommended to avoid injury to conduction system. Normally right judkin catheter is used to facilitate crossing the defect but in certain situations cut pigtail can be improvised for crossing. Once the defect is crossed the wire may be parked in left pulmonary artery (LPA) (fig-2).

But for the ease of snaring, the terumo wire may be parked in superior or inferior vena cava with some effort. Once AV loop is established, appropriate sheath is introduced and parked in transverse arch. LV angiogram can be repeated at this point to confirm size of the defect (fig-3).

Preloaded nit-occlud® PDA-R device is advanced through the sheath while carefully advancing wire attached ahead of device which is part of the attaching equipment. The distal part of the device is released and can be easily seen on

fluoroscopy by two dots as a guide for deployment. The whole system is pulled back without releasing the proximal end till the dital end is approximated to the LV side of the defect guided by check angiograms via pigtail (fig-4).

Figure-1: Nit-occlud® PDA-R (pfm medical) device which was used for closure of doubly committed subarterial ventricular septal defect.

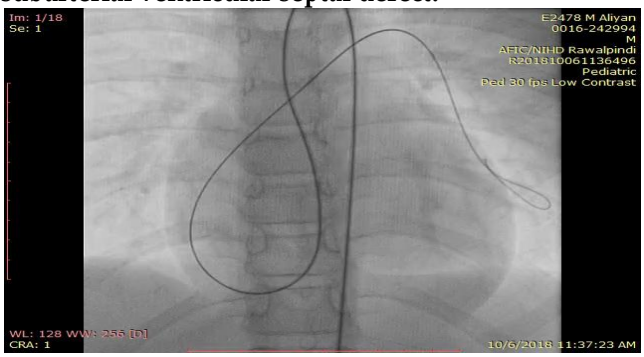

Figure-2: After crossing the defect, the wire is parked in LPA.

Figure-3: LV angiogram done while sheath is parked in aorta to estimate the size of VSD.

Once the LV side position is established, the proximal part is released. Check angiograms in LAO 90 (fig-5).

LAO 25, Cranial 25 (fig-6) and RAO 30 degrees is done to confirm position of device. Best position to confirm the device seems to be LAO 90. When device is released in this position, there is horizontal tilting confirming more accurate position. The pigtail is never pulled back to aorta without wire as the catheter may entangle

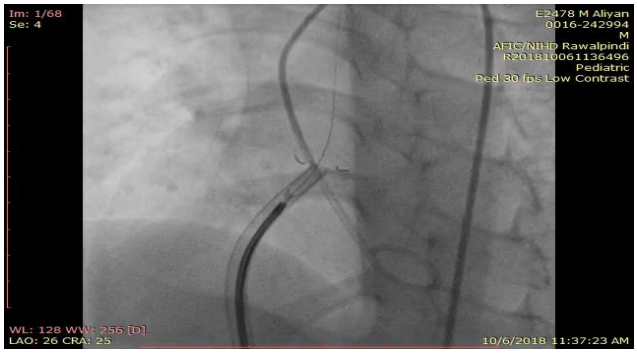

Figure-4: Distal end of device is approximated to the LV side of the defect guided by check angiograms.

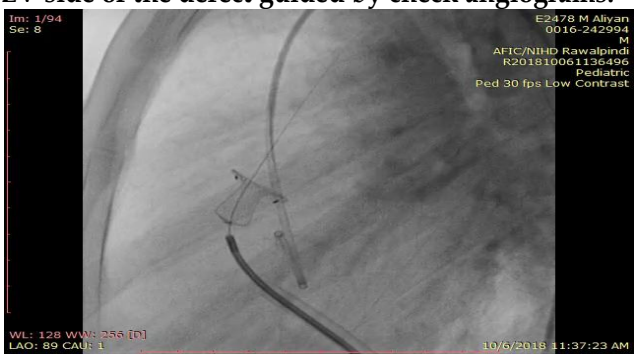

Figure-5: Position of device before release in LAO 89.

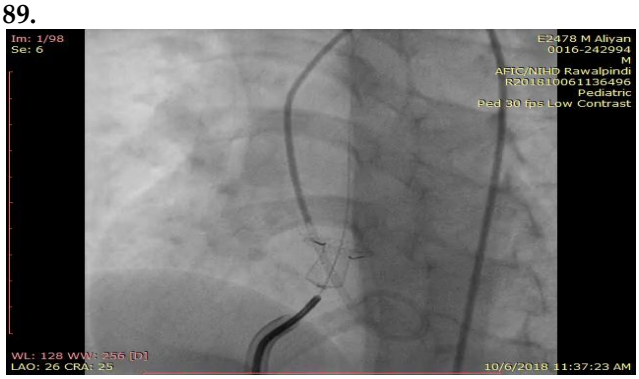

Figure-6: Position of device before release in LAO 26, Cranial 25.

with the device and pull it causing embolization. Post procedure the femoral artery pulse is ensured serially till peripheral pulses are patent. Otherwise proper anticoagulation is indicated with heparin or streptokinase when thrombosis is suspected. In rare situations when above measu-

res fail to resolve thrombosis, tissue plasminogen activator (TPA) administration or vascular surgeon consultation may be requested. Detailed echocardiogram is done next morning before discharge and femoral pulse is ensured. Follow up visits are advised after 2 weeks, 6 months, then yearly.

RESULTS

All cases showed complete closure of the defect with no residual flow. Estimated pulmonary artery pressures on echocardiogram resolved to normal gradually. Adverse events noticed were transient 2:1 heart block in one patient during the procedure while crossing the defect and snaring the wire. The block settled gradually at the end of the procedure. In another patient it was difficult crossing the defect and cut pigtail-catheter was used in LAO 90 projection, the process of crossing took about 25 minutes, rest of the procedure was uneventful. Although this was small number of patients, complications may be anticipated at each step and trouble shootings should be known or consulted.

Table: Sizes of Nit-Occlud® PDA-R used in patients.

No. of Devices	Stent	Disc	Length	Sheath
	4mm	8mm	6.5mm	6F
	5.5mm	10mm	7mm	6F
2.	7mm	12mm	8.5mm	6F
2.	8.5mm	14mm	9.5mm	6F
2.	10mm	16mm	11mm	8F
	11.5mm	18mm	12mm	8F

Mean age was 6 years. Nit-occlud® PDA-R devices used ranged from 8, 4 to 18, 11.5 (table). The narrowest point of VSD doubled for the selected device was found to be effective sizing. Postoperative Echocardiography revealed neither residual VSD nor significant pulmonary regurgitation. No fatality was observed. Various size of devices were used (table).

DISCUSSION

This procedure is unique in the sense that it was done for the first time in our country Pakistan at armed forces institute of cardiology. Although this type of VSD is common among VSDs and all were used to be closed by surgery[12]. Transcatheter device closure of DCSAVSD is relatively new technique saving the time, complications, hospital stay, economics and cosmetics[13]. If performed carefully with proper indications and appropriately selected device, it saves many patients from morbidities and mortalities. Surgical backup is mandatory to assist in case of implantation failure or embolization. Spontaneous closure is rare[4]. This type of VSD is complicated due to coronary cusp prolapse and associated aortic incompetence. In about 39% of the patients the aortic valve is involved[2]. Our cases showed cusp prolapse in about 40% of population and 10% of them indicated valve regurgitation. But this was small patients' number and results may vary in large sample. Valve incompetence in this VSD is an acquired lesion if left untreated for later age[6].

Device closure of DCSAVSD may be carried out as early as 2.5 years of age to avoid further associated complication of cusp prolapse and valve regurgitation[18]. Softconsistency of the device may be non-damaging to valve leaflets and minimize regurgitation due to leaflet approximation by the device disc. However, nitocclud VSD coil may be used instead for packing the defect and lifting the leaflets for approximation.

The older is the patient having valve incompetence, the disease is gradually escalating[7].

Trivial or mild aortic valve regurgitation along with DCSAVSD may be subjected to device closure and prolapse may be abolished but more regurgitation mandates surgical closure of VSD along with valve repair/replacement.

All these types of VSDs should be closed irrespective of cusp prolapse or valve regurgitation. Closureof this VSD not only closes the defect and avoids valve prolapse but also prevents RVOT obstruction and damage to pulmonary valve[9]. Whenever slightest of regurgitation is noticed along with DCSAVSD on echocardiogram, it should be closed at the earliest[10].

Various innovations may be adopted for transcatheter closure of this defect[11]. Another device currently used for VSD closure is Konar-MF VSD occlude (Lifetech). It is anticipated that this device may prove useful for DCSAVSD closure with reasonable results due to very soft consistency and benign during negotiation with the valve leaflets. Additionally, this device does not need AV loop and avoids snaring at the cost of arterial line on both sides. During closure of this defect electrophysiologist should also be kept on board to deal with possible arrhythmias.

We have selected many other patients after this initial experience and planto close more defects which was not possible in the past without surgery. Due to innovations and variety of devices used for closure, we are trying to economize the total cost in the face of economic hardships currently in our country.

It is therefore recommended that all DCSAVSDs should be thoroughly screened by echocardiogram to asses the suitability of device closure. VSDs suitable for device closure should be closed at the earliest to avoid further complication.

CONCLUSION

Due to high prevalence of DCSAVSD in Asian population compared to western world, device closure is safe and economical procedure to avoid further morbidities of valve incom-petence and surgical scar. Device selection against the defect nature is key for success in this scenario.

CONFLICT OF INTEREST

This study has no conflict of interest to be declared by any author.

REFERENCES

1. Devlin PJ, Russell HM, Monge MC, Patel A, Costello JM, Spicer DE, et al. Doubly committed and juxta arterial ventricular septal defect: outcomes of the aortic and pulmonary valves. Ann Thorac Surg 2014; 97(6): 2134-41.
2. Tatsuno K, Ando M, Takao A, Hatsune K, Konno S. Diagnostic importance of aortography in conal ventricular-septal defect. Am Heart J 1975; 89(2): 171-77.
3. Anderson RH, Wilcox BR. The surgical anatomy of ventricular septal defects associated with overriding valvar orifices. J Card Surg 1993; 8(2): 130-42.
4. Gittenberger-De Groot AC, Poelmann RE. Normal and abnormal cardiac development. Moller JH, Hoffman JI, eds. Pediatric Cardiovascular Medicine. New York, NY: Churchill Livingstone 2000; 3-20.
5. Yip WC, Ho TF, Chan KY, Lee CN, Ong KK, Koh BC. Comparison of spontaneous closure rate in children with different types of ventricular septal defects. J Paed Child Health 1997; 33(suppl-1): S41-S45
6. Wei XZ, Kang SZ, Xiang DW. Surgical repair of ventricular septal defect in 2085 pediatric patients. Asian Cardiovasc Thorac Ann 1997; 5(2): 83-86.
7. Griffin ML, Sullivan ID, Anderson RH, Macartney FJ. Doubly committed subarterial ventricular septaldefect: New morphological criteria with echocardiographic and angiocardiographic correlation. Heart 1988; 59(4): 474-79.
8. Mori K, Matsuoka S, Tatara K, Hayabuchi Y, Nii M, Kuroda Y. Echocardiographic evaluation of the development of aortic valve prolapse in supracristal ventricular septal defect. Eur J Pediatr 1995; 154(3): 176-81.
9. Momma K, Ando M, Matsuoka R, Joo K. Interruption of the aortic arch associated with deletion of chromosome 22q11 is associated with a subarterial and doubly committed ventricular septal defect in Japanese patients. Cardiol Young 1999 9(5): 463-67.
10. Yacoub MH, Khan H, Stavri G, Shinebourne E, Radley-Smith, R. Anatomic correction of the syndrome of prolapsing right coronary aortic cusp, dilation of the sinus of Valsalva, and ventricular septal defect. J Thorac Cardiovasc Surg 1997; 113(1): 253-61.
11. Van Praagh R, McNamara JJ. Anatomic types of ventricular septal defect with aortic insufficiency: diagnostic and surgical considerations. Am Heart J 1968; 75(5): 604-19.
12. Tatsuno K, Konno S, Ando M, Sakakibara, S. Pathogenic mechanisms of prolapsing aortic valve and aortic regurgitation associated with ventricular septal defect. Circulation 1973; 48(5): 1028-37.
13. Tweddell JS, Pelech AN, Frommelt PC. Ventricular septal defect and aortic valve regurgitation: pathophysiology and indications for surgery. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2006; 9(1): 147–52.
14. De Leval MR, Pozzi M, Starnes V. Surgical management of doubly committed subarterial ventricular septal defects. Circulation 1988; 78(1): III40–III46.
15. Moreno-Cabrol RJ, Mamiya RT, Nakamura FF, Brainard SC, and McNamara JJ. Ventricular septal defect and aortic insufficiency: surgical treatment. J Thorac Cardiovasc Surg 1977; 73: 358–365.
16. Chauvaud S, Serraf A, Mihaileanu S. Ventricular septal defect associated with aortic valve incompetence: results of two surgical managements. Ann Thorac Surg 1990; 49(6): 875-80.
17. Bonhoeffer P, Fabbrocini M, Lecompte Y. Infundibular septal defect with severe aortic regurgitation: a new surgical approach. Ann Thorac Surg 1992; 53(5): 851-53.
18. Lincoln C, Jamieson S, Joseph M, Shinebourne E, Anderson RH. Trans-atrial repair of ventricular septal defects with reference to their anatomic classification. Surg 1977; 74(1): 183–90.