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Submission date: 20-Mar-2021 10:53AM (UTC+0800)
Submission ID: 1537530382
File name: rial_doubly_commited_ventricular_septal_defect_A_case_report.pdf (834.71K)
Word count: 3390
Character count: 18217

Sapporo Medical Journal
Volume 54, Issue 08, August, 2020

Transcatheter Closure of Subarterial Doubly Commited Ventricular Septal Defect : A Case Report

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Abstract— Ventricular septal defects (VSDs) are a common congenital heart disease (approximately 20%). The most common are the perimembranous VSD (around 70%), while completely muscular VSD may occur around 15% of the cases. Supracrystal defects are quite rare accounting for 5% of all VSD. Indications to VSD closure are symptoms of heart failure, signs of left heart chambers overload, and history of endocarditis. The traditional treatment is surgical repair. The surgical approach is considered to be the gold standard, but it is associated with morbidity and mortality, patient discomfort, sternotomy and skin scar. Percutaneous techniques have been developed in order to reduce the impact of such drawback of surgery. Since the first VSD closed by a transcatheter approach by Lock et al. various techniques have been used. Closure of VSD using Ampatzeroccluders has been greatly improved and widely reported. VSD inselected patients may be closed percutaneously using an Amplatzer Ductal Occluder II (ADO II) device, as an off-label therapy. It appears that ADO II may be the preferable device for the closure of defects of moderate size (2-5 mm), especially in infants and small children, because of its better profile.

Keywords— Ventricular septal defect, Transcatheter closure, ADO II

1. Introduction

Ventricular septal defect (VSD) is the most common congenital abnormality. Subarterial doubly committed (SADC) VSD is a unique type of VSD located just below the aortic and pulmonary valves, accounting for about 5% of all VSDs. Due to its unique location, this VSD is characterized by a low tendency to close spontaneously and aortic valve prolapse or progressive insufficiency due to the "Venturi effect" of the left to right shunt. Open-heart surgery is said to be the standard standard for VSD SADC therapy, but this method is associated with complications of cardiopulmonary bypass such as systemic inflammatory response, myocardial dysfunction, and blood transfusions, especially for pediatric patients.[1] In the last two decades, percutaneous approaches to VSD closure have been developed. Several tools have been used for this VSD closure.Here we report a case of a patient with VSD SADC who had VSD closure with a transcatheter using Amplatzer Ductal Occluder II.

2. Case

An. R, 2 years old, with a body weight of 13 kg, has complaints of rapid fatigue and tightness when do activity. There was no previous blue history. The patient was known to have heart disease from birth.From physical examination, it was found that the general condition was sufficient with a pulse of 130 x/minute, breathing 24 x / minute, a temperature of 36ºC. Head and neck physical examination was normal, there was no increase in JVP, chest examination found ictus cordis in the left ICS V midclavicular line, auscultation of a single S1 S2 heart, no gallop and extrasystole, found systolic murmur in ICS III parasternal line left grade III / VI. Lung auscultation heard vesicular breath sounds throughout the lung fields, no rhonchi or wheezing was heard. From the examination of the abdomen and extremities within normal limits.Then performed an X-ray examination with cardiomegaly with a cardiothoracic ratio of 58% and both lung fields were within normal limits (Figure 1.)

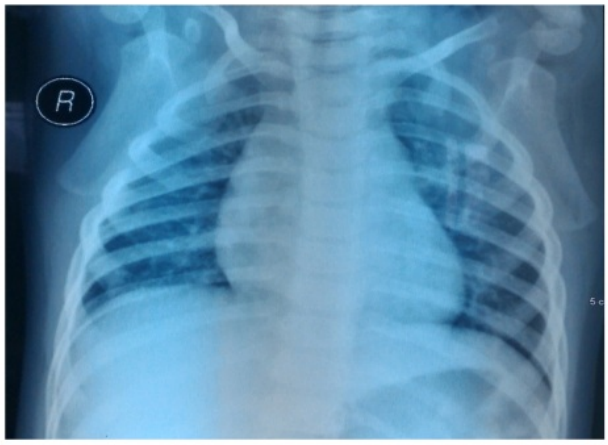

Figure 1. Chest X-ray examination

Laboratory tests showed Hb 13.7g/dL, leukocytes 14.050/uL, platelet 314,000/uL, BUN 10 mg/dL, serum creatinine 0.93 mg/dL, albumin 4.4 g/dL, SGOT 44 U/L, SGPT 23 U/L, potassium 4.4 mmol/L, sodium 139 mmol/L, chloride 110 mmol/L, APTT 26.8 seconds, PPT 10 seconds. Echocardiography showedsolitus site, AV-VA concordance, normal pulmonary vein drainage, heart chambers LA-LV was dilated, valves were within normal limits, atrial septum was normal, ventricular septum appears to be VSD subarterial doubly committed moderate size 3.1 - 4.5 mm, no patent ductus arteriosus, normal left aortic arch, normal left ventricular systolic function. The conclusion of echocardiography was that VSD SADC was being L to R shunt (Figure 2.).

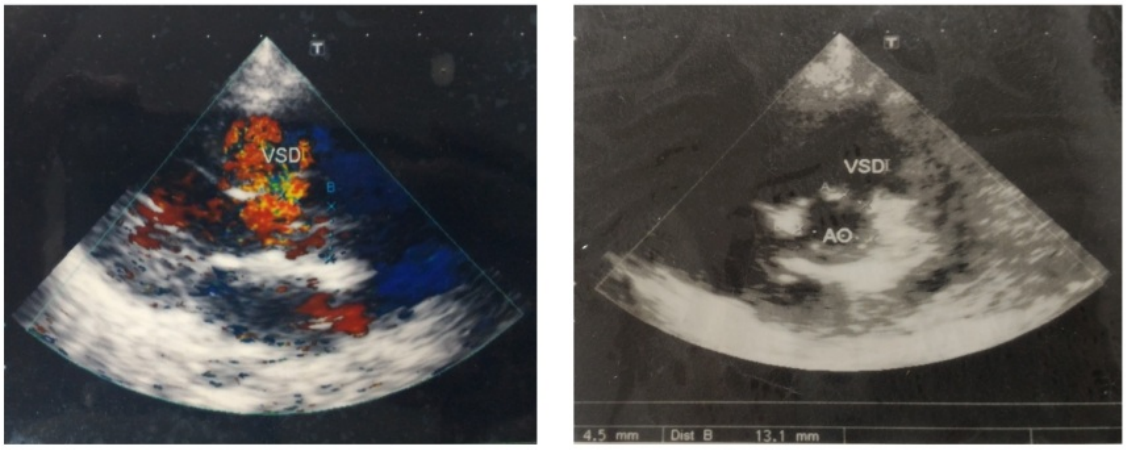

Figure 2. VSD view on echocardiography examination

After the diagnosis of VSD SADC was established, the patient was planned to undergo percutaneous closure of the VSD. The procedure begins by inserting a 5F pigtail catheter through a sheath in the right femoral artery into the aorta with the help of a wire, then pushing it into the LV. Then a left ventriculography was performed to see the contrast flow from the left ventricle to the right ventricle. The results of this left ventriculography show the presence of contrast flow from the left ventricle to the right ventricle via VSD SADC, then measurement of the interventricular septal defect. After that, the pigtail catheter was removed. In addition to measurements using left ventriculography, measurements were taken using echocardiography before the procedure. The result was a ventricular septal defect measuring 5 mm (Figure 3.).

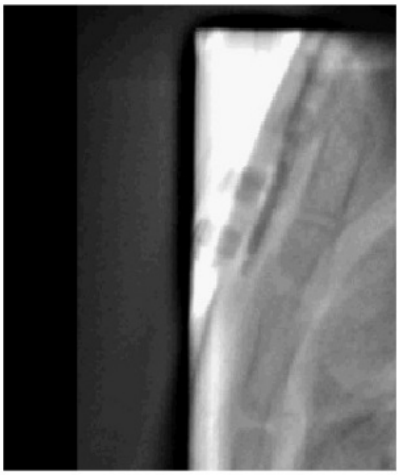

Figure 3. Left ventriculography

Then a JR 4F catheter using a long wire 0.035 Terumo 260 cm was inserted through the sheath in the right femoral artery to the left ventricle, then through the VSD to the right ventricle then to the pulmonary artery. After that, the MPA catheter was inserted through the sheath in the right femoral vein with the help of a wire to the inferior vein cave, then the right atrium, right ventricle, to the pulmonary artery. Then insert the 10F snare kit through the MPA catheter into the pulmonary artery. The long wire in the pulmonary artery was successfully pulled with a snare kit and then pulled into the right ventricle and right atrium until it came out through the right femoral vein. After that, the sheath and catheter were removed.

The next procedure was followed by inserting the Mullin Delivery System SFP 5F through the right femoral vein with the help of a wire to the right atrium, then the right ventricle, then through the VSD. Then the Amplatzer Ductal Occluder (ADO) AGA 0604 was inserted and positioned on the VSD. When ADO enters the left ventricle, it was developed distally and then it was pulled proximal to the waist, after which the proximal part was developed (Figure 4.).

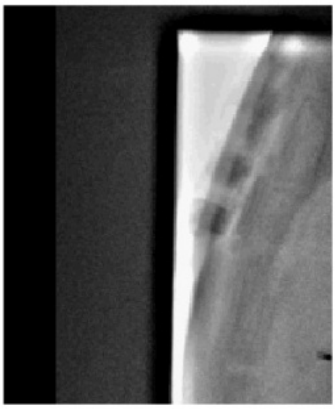

Figure 4. After installing ADO

After the procedure was completed, evaluated by echocardiography and left ventriculography with the results of the ADO position wasin correct position, no residual flow was seen (Figure 5.). After the action the patient was given aspirin at a dose of 50 mg / day for 6 months.

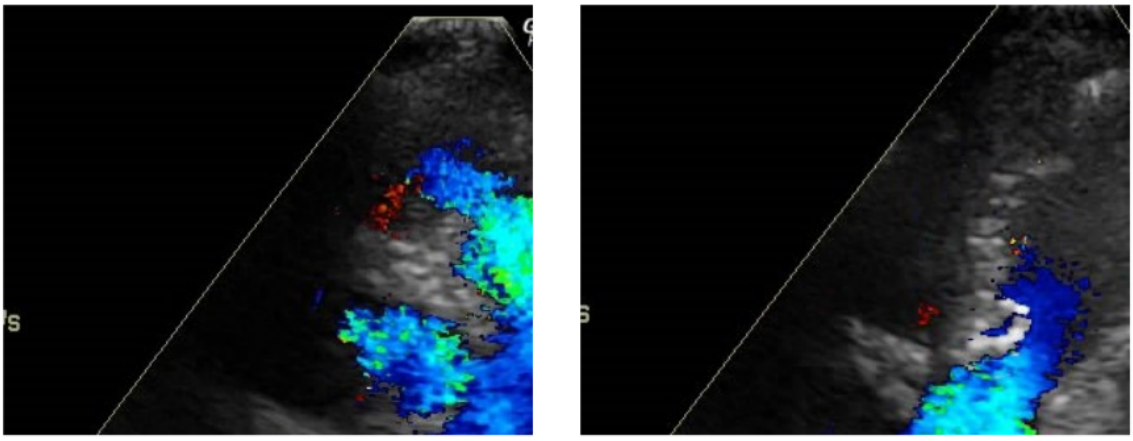

Figure 5. Echocardiographic examination after ADO placement

4. Discussion

Ventricular septal defect is the most common congenital heart defect, accounting for about 20% of total congenital abnormalities. VSD is a developmental disorder of the interventricular septum resulting from deficiency of growth or failure of alignment or fusion of the ventricular septal components.[2] The ventricular septum is divided into small membranous sections and large muscular sections. The muscular part consists of the inlet septum, trabecular septum, and outlet septum. VSD can be obtained in any part of the interventricular septum. Approximately 70-80% of defects are perimembranous where 5-20% are located in the muscular part. Outlet defects (infundibular, supracristal, subpulmonary, or doubly committed arterial) account for 5–7% of total VSDs. This defect is located in the outlet (conal) septum and part of the rim is formed by the aortic and pulmonary annulus. The aortic valve leaf can prolapse via VSD and cause aortic valve insufficiency. The VSD inlet (AV channel) accounts for about 5-8% of the total VSD. This defect is located posterior and inferior to the perimembranous defect under the septal valve of the tricuspid. This type of VSD cannot be used for transcatheter closure because there is no supporting tissue between the defect boundary and the atrioventricular valve network. Muscular or trabecular defects account for about 5-20% of total VSDs. These defects are often multiple. The type of "Swiss cheese", namely multiple muscular defects, is very difficult to close surgically.[3] This patient had a subarterial doubly committed type VSD (SADC) measuring about 3.1 - 4.5 mm.

The progression of VSD has a wide spectrum, from spontaneous closure to heart failure and mortality during infancy. Spontaneous closure usually occurs in childhood, usually up to 2 years of age. this closure is often seen in muscular defects (80%) followed by perimembranous defects (35-40%). Outlet defects have a small incidence of spontaneous closure, and inlet defects cannot close spontaneously.[2] Patients with a small VSD, the patient can remain asymptomatic with normal growth and development. Patients with moderate to large VSD sometimes have complaints of stunted growth and development, decreased activity tolerance, recurrent pulmonary infections, and heart failure in infancy. [4]

The course of VSD is also sometimes accompanied by several complications. Aortic valve leaf prolapse usually occurs in SADC VSDs and less frequently in perimembranous VSDs. Secondary aortic insufficiency is associated with aortic valve leaf prolapse. This complication occurs in only about 5% of patients with VSD. Oarta regurgitation occurs because the leaf of the right aortic valve does not support properly combined with the Venturi effect caused by the VSD jet which then results in prolapse of the valve leaf. Aortic regurgitation is progressive in its course and the presence of aortic regurgitation even though it is mild or aortic valve prolapse without aortic regurgitation is an indication for closure. [2]

Children with excess volume in the atrium and left ventricle due to VSD require VSD closure to prevent pulmonary hypertension, ventricular dilation, arrhythmias, aortic regurgitation, and the risk of endocarditis.[5] Surgery is the standard procedure for closure of VSDs. Although this surgical procedure is

considered safe, it carries risks including a complete AV block of 1-5%, a significant residual VSD of 1-10%, a need for re-operation of 2%, and even 0.6-5% of death. In addition, infections, tachyarrhythmias, and neurological complications, patient discomfort, sternotomy, and skin scarring can also occur.[6,7]

In the last two decades, percutaneous approaches have been developed for closure of VSD. This percutaneous approach is preferred by patients and families because of its less psychological effects (no scarring after the procedure), less hospitalization time, less discomfort and pain, and does not require hospitalization in the intensive care unit.[8] Since the first time VSD was closed with a transcatheter approach by Lock and friends, several tools have been used for VSD closure, including Rashkind double umbrella, CardioSEAL (NMT Medical, Boston, MA, USA), STARFlex (NMT Medical, Boston, MA, USA), coils, Amplatzer Muscular and Membranous VSD, Amplatzer Septal Occluder, Amplatzer Duct Occluder, and Duct Occluder II (St. Jude Medical, Inc. St. Paul, Minnesota, USA), and Chinese symmetrical and asymmetrical occluders (Shanghai Memory Alloy Materials Co., Ltd, China and Huayishengjie Medical Corp., Beijing, China) which is a variation of the Amplatzer device.[3,9]In this patient, Amplatzer Duct Occluder II (ADO II) was used for percutaneous closure of VSD. The action went well, there were no complications. The tool is in a good position and no residual flow is obtained.

Rashkind double umbrella is the first tool used for closing VSDs. This device is a single-disk consisting of polyurethane foam with a hexagonal shape. This tool was originally designed for closure of PDA or ASD. 1988 Lock and colleagues first reported its use for closure of VSD.[9] After that many reports about the results of using this Rashkind. These early devices and systems achieved moderate levels of success, where residual flow was obtained, inducing valve regurgitation (aortic and tricuspid valves), arrhythmias, and bleeding, as well as difficult instrument placement.[8]

Currently, there are many reports of closure atrial septal defect (ASD), patent ductus arteriosus (PDA), VSD using Amplatzeroccluder. The use of Amplatzer for VSD closure was first performed by Lee and colleagues who succeeded in inserting an Amplatzer septal occluder in a patient aged 50 years with acute VSD post myocardial infarction. patients have good clinical development and have a small residual flow.[10] VSDs in selected patients can be closed percutaneously using the ADO II device, as off-label therapy. ADO II is preferred for defects of medium size (2-5 mm), especially for infants or young children, because of its better profile. [8]

ADO II is made of Nitinol wire mesh which is layered, flexible and connected to the disk at each end. Hemostasis results from multiple layers of wire mesh. The fabric-free technology makes it easier to follow the angulation. The delivery system required is only 4 or 5 France in size. Therefore, using ADO II simplifies the procedure, improves the success rate of insertion, and reduces the rate of complications related to vascular access. This device has a tendency to elongate when tension is obtained when pulling the cable and has a longer central waist which prevents mechanical compression on the perinodal network. Research conducted by Kanaan and colleagues in 2015 regarding the follow-up of VSD patients who were closed percutaneously using ADO II showed satisfactory results. The procedure was performed successfully in 29 of 31 patients (93.5%) without any significant complications. There was no post-procedure aortic or tricuspid valve regurgitation. The residual VSD flow was obtained immediately after implantation of the device, but disappeared in a median follow-up period of 38 months in 27 of 31 patients. There was no incidence of AV block or conduction abnormalities during implantation or follow-up. [8]

The results of transcatheter VSD closure have been reported by one of them, namely the European VSD registry. Reported from 430 patients consisting of 119 muscular, 250 membranous, 16 multiple, and 45 postoperative residual VSDs. The tools used include Membranous Amplatzer (213 tools), Muscular Amplatzer (151 tools), PDA Amplatzer (12 tools), ASD Amplatzer (7 tools), Starflex (7 tools), Coil (9 tools). The procedure's success rate is 95.3%. There was an initial complication rate in 55 patients (12.7%) with significant complications in 28 patients (6.5%). There was one death (0.2%) with vascular complications of 0.7%, hemolysis of 1.2%, infection of 0.5%, and appliance embolization of 0.9%. Tachyarrhythmias occurred in 3 patients (0.7%) with incidence of total AV block in 12 patients (2.8%). Six of these patients developed a total AV block at the time of the procedure, and 6 patients developed a total

AV block within 1 week after the procedure. During 4-18 months follow-up, 4 patients (0.9%) had total AV block and all of these patients were taking AmplatzerPerimembranous VSD (Carminati M, et al., 2007). Research Holzer and colleagues also reported the results of closure of perimembranous VSD using AmplatzerPerimembranous VSD in 100 patients weighing more than 5 kg. The procedure success rate was 93 patients (93%). Complications occurred in 29 patients (29%) where the most common complication was arrhythmia (13%). In addition, there was new or increased aortic valve regurgitation in 9.2% of patients where this regurgitation disappeared at follow-up in 4 patients and was trivial or mild in 4 patients. New or increased tricuspid valve regurgitation also occurred in 9.2% of patients where this regurgitation disappeared at follow-up in 3 patients and was trivial or mild in 5 patients. Only 1 patient had moderate or worsening aortic or tricuspid valve regurgitation during a median follow-up of 182 days. [11]

Complications of appliance embolization can be prevented by selecting the appropriate size. The size of the tool used should be based on the size of the defect plus 0.5-1 mm. [8] Another complication that still often occurs is total AV block. This complication appears to be more common in younger sufferers. Compared with surgery, where total AV block appears in the early phase after surgery, in patients who have percutaneous closure, the appearance is unpredictable and can appear for a long time. [7]Possible mechanisms, namely mechanical compression, tissue, or inflammatory reactions are likely to resolve in the early phase. The formation of scar tissue in the conduction tissue and chronic inflammation may be the cause of the appearance of AV block in the late phase. Some cases spontaneously returned to sinus rhythm, or with temporary pacemakers, corticosteroids, and / or salicylic acid therapy, or device removal. [12] The use of ADO II may reduce the incidence of total AV block due to the flexibility of the tool or the ease of the implantation process. [8]

VSD closure by means can also be done with a perventricular approach. This technique is used as an alternative to closing VSD SADC. This technique is performed surgically with 2 types of incisions, namely sternotomy 3-4 cm in the lower median or mini-thoracotomy 2-3 cm in the anterior left. The operator then inserts the needle into the right ventricular space with the help of transesophageal echocardiography (TEE), followed by a guidewire and a double-lumen delivery sheath. In a study conducted by Zhang and colleagues, an eccentric enclosure device 1-2mm larger than the size of the VSD was used. [1]

5. Conclusion

Ventricular septal defect (VSD) is the most common congenital abnormality. Approximately 70-80% of defects are perimembranous, followed by muscular or trabecular defects in 5-20%, inlet VSD (AV canal) in 5-8%, and outlet defects (infundibular, supracristal, subpulmonary, or doubly committed arterial) around 5-7% of the total VSD. Surgery is the standard procedure for closure of VSDs. Although this surgical procedure is considered safe, it carries many risks. Many percutaneous approaches have been developed for VSD closure. One that has been widely reported is using Amplatzer. The Ductal Occluder II (ADO II) Amplatzer device can be used for VSD closure as an off-label therapy. ADO II can be an option for medium-sized (2-5 mm) VSDs because of the advantages of a better profile and easier angulation. We report a case of a patient with VSD SADC measuring approximately 3.1 - 4.5 cm who successfully underwent transcatheter VSD closure using the ADO II device, and had no complications.

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PAGE 1

PAGE 2

PAGE 3

PAGE 4

PAGE 5

PAGE 6

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