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Original Article

Muscular ventricular septal defect closure with Gianturco coil at Soetomo hospital (a case report)

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Ventricular septal defect (VSD) is the most common congenital heart disease (CHD) in children.[1,2] It occurs in 1.5 to 3.5 of 1,000 live births and constitutes 20% of congenital cardiac defects.[1] The VSD may be small, medium or large and is classified based on its location in the interventricular septum. There are four types of VSD, i.e., perimembranous (80% of VSDs), muscular type (5% to 20%) inlet or AV canal type (8%), and finally, subpulmonary (5% to 7%).[1-4] When multiple muscular defects are seen, it is often referred to as "Swiss-cheese" type of VSD. [1,2]

The management strategies, which consist of medical, surgical and intervention techniques, depend to a large degree on the size of the VSD.[1-4] Approximately 40% of VSDs spontaneously and completely closed, with closure rates approaching 80-90% by age 2 years.[2] Indication of VSD closure are symptoms of heart failure, left heart chambers overload and history of endocarditis. The surgical approach is considered gold standard but it is associated with morbidity and mortality, high cost, patient discomfort, sternostomy and skin scar.[3] Since 1988, percutaneous techniques have been conducted in order to reduce those drawbacks of surgery. More recently, percutaneous techniques and devices have been developed specifically for closure of muscular VSD (mVSD) and perimembranous VSD (pmVSD) using either the Rashkind double umbrella, the Bard Clamshell, the Button device, the Amplatzer septal, duct or muscular VSD occluder, or the Gianturco coils.[3,4]

Gianturco coils have been widely used to close unwanted vascular communications and small- to moderate-sized patent ductus arteriosus, with excellent closure rates. In 1999, Latiff et al successfully used this coil to close multiple muscular VSDs in a 10-month old boy. Thus, percutaneuos closure of VSDs using Gianturco coils is a feasible, reasonable alternative to surgery.[5,6] We report a case of four-year-old girl with muscular VSD who underwent cardiac catheterization and transcatheter closure with Gianturo coil in Dr. Soetomo Hospital, Surabaya.

The Case

A 4-year old girl was referred to the Cardiology Outpatient Clinic Soetomo Hospital. The parents noticed their daughter's heart disorder since July

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2008, when she was hospitalized in Sorong. She was then taken by her parents to Dr. Soetomo Hospital for further management. The patient had never any episodes of cyanosis, dyspnea, or edema. The oral intake was adequate.

She was delivered spontaneously at term by a healthy mother. Her birth weight was 2750 grams. She was breastfed until 6 months old. Her immunization status was up dated. The growth and developmental pattern was normal. Examination disclosed an alert girl weighing 18 kg, with heart rate 100 beats per minute, respiratory rate 30 times per minute and temperature 36.8°C. There were no pale, dyspnea, or cyanosis found. The jugular venous pressure was normal. There was a grade III/VI early systolic murmur at the intercostal III – IV left parasternal line. There were no hepatomegaly, edema, or other signs of cardiac failure. The capillary refill time was less than 2 seconds, and there were no clubbed fingers. Other findings were normal. The chest roentgenogram showed normal size and shape heart. The cardio-thoracic ratio was 54% with normal lung vascularization. Echocardiography revealed normal chamber and valve. A small trabecular muscular ventricular septal defect was found with the diameter of 2.5 mm. Systolic function of the left ventricle was normal, with ejection fraction 64%. Doppler showed normal pulmonary artery (PA) flow and left to right shunt at the VSD.

Laboratory examinations were taken for the preparation of the cardiac catheterization showed normal values. Informed consent for the procedure was obtained from the parents.

On the 2nd day of hospitalization, the patient underwent cardiac catheterization and transcatheter closure of the marginal muscular VSD. Antibiotic cefazoline 700 mg was given intravenously 30 minutes before the procedure. General anesthesia was performed during the procedure. The pulmonary artery pressure was 25/15 mmHg (mean pressure 20 mmHg), and the aortic pressure was 120/80 mmHg (mean pressure 93.3 mmHg). Angiography of the left ventricle, which was performed during the procedure showed positive contrast filling and normal chamber.

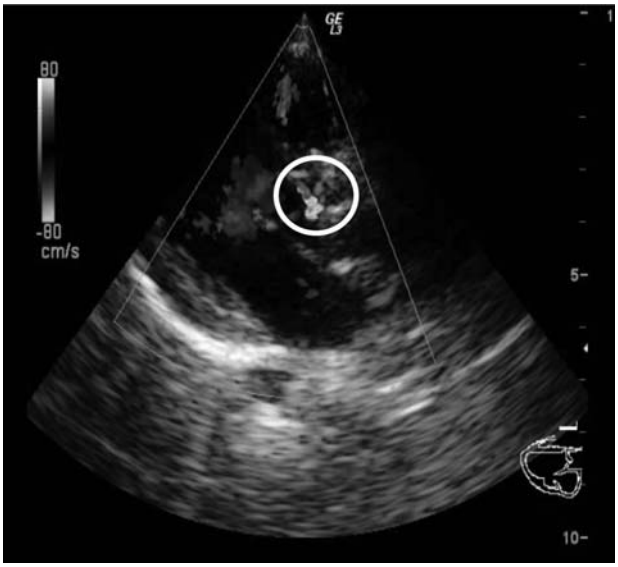

Figure 1. Echocardiography pre catheterization showed small trabecullar muscular VSD.

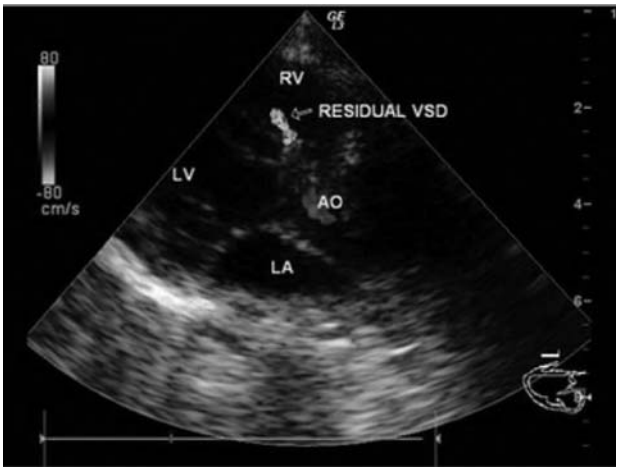

Figure 2. Post-procedure echocardiography showed coil in situ and small residual VSD.

Table 1. Catheterization result

SITE	PHASIC (mmHg)	MEAN (mmHg)	SATURATION (%)	NOTE
LV	120/10	46.6	94.3	
Ao	120/80	93.3	94.3	
RV	25/5	11,6	85.1	Step up Saturation in RV 16,8%
PA	25/15	20	84.4	
RA		5	72.8	
IVC			75.5	
SVC			68.1	

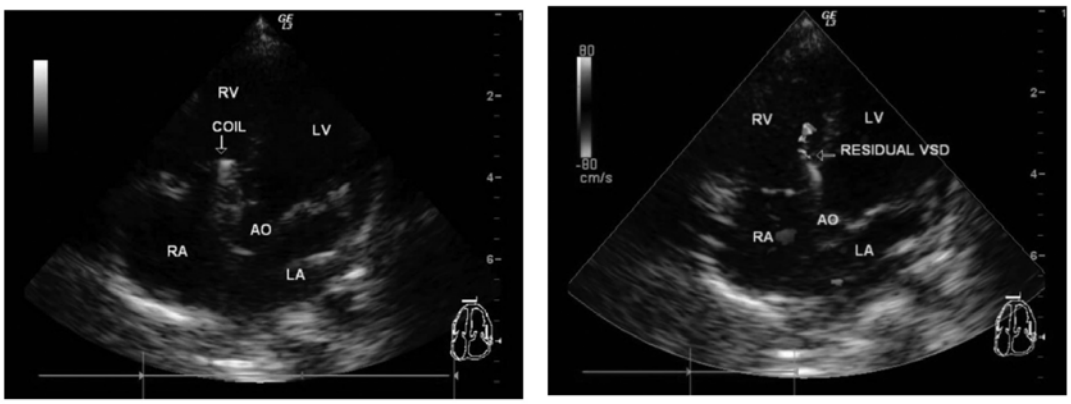

Figure 3. Echocardiography 2 weeks post catheterization showed coil in situ, small residual VSD and trivial AR.

Muscular VSD was found with the diameter of 2.88 mm. After the defect was identified, Amplatzer as the device for closure was ready to be used, but the Mulin catheter was failed to enter the defect because of its small diameter. The operator decided to use other device (coil) which was considered suitable with the size of the defect. A 0.035-inch Straight wire was used in an attempt to probe the VSD from the left ventricular side for coil implantation. An 8 mm Gianturco coil was then used to close the defect. Angiography of the left ventricle after coil implantation showed no residual VSD. There were no hemodynamic problem or other complications observed during the procedure.

Three hours after the procedure, the patient was sent back to the pediatric ward with blood pressure 100/60 mmHg, pulse rate 100 times per minute and respiratory rate 28 times per minute. There was no hematoma or bleeding found. Both dorsal pedis artery showed the same pulse rate. Antibiotic cefazoline was given intravenously 6 and 12 hours after catheterization. Aspirin 80 mg once daily orally was administered.

On the next day, there were no complaints found in the patient. She was able to stand and walk by herself. The oral intake was adequate. The echocardiogram showed coil *in situ* with small residual VSD. The valve function was normal and there was no pericardial effusion detected. On the same day, the patient was discharged in a good condition and planned for follow-up 1, 6 and 12 months after the procedure.

Two weeks after the coil implantation, the patient made a follow-up visit. The patient showed good general condition and had no complaint. The blood pressure was 100/60 mmHg, with pulse rate 100 times per minute and respiratory rate 28 times per minute. From chest auscultation, there was systolic murmur heard at intercostal III – IV grade II/VI left PSL. The echocardiogram revealed coil *in situ*. There were also small residual VSD and trivial AR (aortic regurgitation) detected. Other valve functions were normal and there was no pericardial effusion observed.

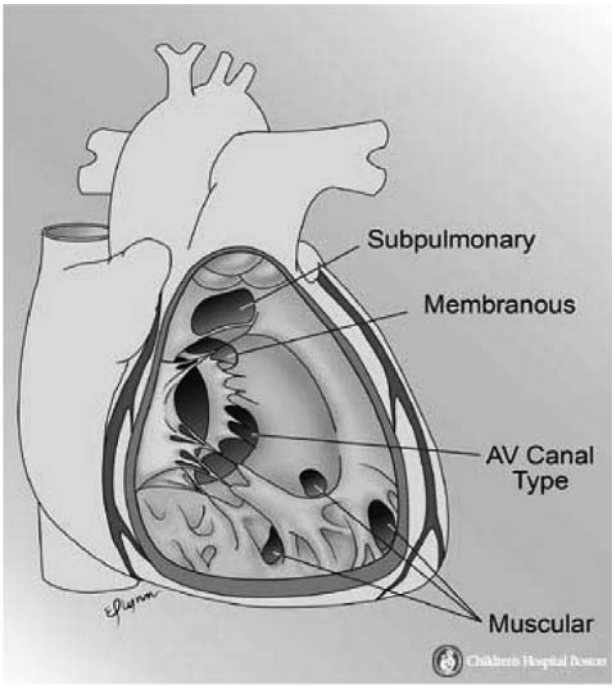

Figure 4. Diagram of types of VSD
Source: Multimedia Library Children Hospital Boston
Available at: http://www.childrenshospital.org/cfapps/mml_index.cfm?CAT=media&MEDIA_ID=356

On 1-month follow-up, the patient came with good general condition. There were no complaints. The patient was conducted chest roentgenogram one month after the procedure and it showed heart with normal size and shape, normal lung vascularization and Gianturco coil *in situ*. The echocardiogram revealed similar results as before.

Discussion

Ventricular septal defect (VSD) is the most common CHD in the first 3 decades of life.[4,6] It occurs when any portion of the ventricular septum does not correctly form or if any of components do not appropriately grow together.[6] VSD is performed as a defect (hole) in the ventricular septum (a dividing wall between the two lower chambers of the heart - the right and left ventricles). Because of this opening, blood from the left ventricle flows back into the right ventricle, due to higher pressure in the left ventricle. This causes an extra volume of blood to be pumped into the lungs by the right ventricle, which can create congestion in the lungs.[6,7]

Ventricular septal defects are typically classified according to the location of the defect in one of the 4 ventricular components: the inlet septum, muscular septum, outlet/infundibular septum, or membranous septum.[1-4,6] Muscular VSD is the second most common type of VSD, occurring in 5-20% of most series. Muscular VSDs are divided into separate distinct regional groups including midmuscular, apical, anterior, and posterior. Midmuscular is the most common subtype of muscular VSD. Defects occurring centrally or along the margin of the interventricular septum and free wall are termed anterior VSDs.[6]

The clinical presentation is largely dependent upon the size of the VSD.[1-4] In small defects, the patients are usually asymptomatic and are detected because a systolic cardiac murmur heard on routine examination.[1-2] In muscular VSDs, the murmur may be best heard over the lower sternal area. In very small defects, the murmur though begins with first heart sound, may not last through the entire systole; the shorter the murmur, the smaller is the defect. The intensity of the murmur may vary between grades II-V/Vl. There is no significant variation of this murmur with respiration. This murmur is produced by flow across the VSD. [2]

The patient in this case, was asymptomatic. She never had any episodes of dyspnea, cyanosis and sweating. The patient also gained weight. The cardiac disorder was first noticed unintentionally when she was hospitalized in Sorong. From physical examination, revealed an early sistolic murmur grade III/VI which was heard best on intercostal III – IV left lower parasternal line.

In case of small VSDs, the chest x-ray will be normal. As the size of the VSD increases, the heart size will enlarge. The electrocardiogram may be normal in very small defects or may show evidence for left ventricular hypertrophy in small to moderate defects while it may show biventricular or right ventricular hypertrophy in moderate to large defects.[1]The diagnosis of VSD can be confirmed using echocardiography. The location and size of the defect can be defined and associated anomalies can be identified. Echocardiogram shows increase in left atrial and left ventricular size, which is again dependent upon the size of the VSD.[2] Cardiac catheterization is rarely necessary for patients with uncomplicated VSD and without evidence for pulmonary hypertension.[1,2,6]

The chest x-ray of the patient in this case, showed normal size and shape of the heart, with the cardio-thoracic ratio 54% and normal pulmonary vascular markings. From the echocardiography revealed normal chamber and valve. A small trabecullar muscular VSD was found with echodrop diameter of 2.5 mm. Doppler showed left to right shunt VSD and normal pulmonary artery. As all of the findings were combined, the diagnosis of small trabecullar muscular VSD was confirmed for the patient.

Morbidity and mortality are influenced by the number and size of VSDs, the degree of left-to-right shunting. Muscular VSDs may spontaneously decrease in size and eventually closed. Small muscular VSDs have the greatest likelihood of spontaneous closure, with closure rates approaching 80-90% by age 2 years. Muscular defects in these patients decrease in size due to growth of the ventricular myocardium, which fills in the defect.[6] If spontaneous closure of a VSD did not occur in childhood or adolescence, the chance that it will be closed in adult life is small.[8] One study that used fetal echocardiography showed that 33% of all defects closed in utero, 44% of defects

spontaneously closed within the first postnatal year, and 23% of defects did not close. Irrespective of the hemodynamic effects of VSDs, there is an additional risk of developing subacute bacterial endocarditis.[9]

The patient in this case, had a muscular VSD with the size of 2.5 mm. which did not reach spontaneuos closure by the age 4 years. At age above 2 years, the chance of VSD reach spontaneous closure was very small. This may bring higher risk of subacute bacterial endocarditis. Thus, it was considered an indication for closure.

Indications to VSD closure are symptoms of heart failure, signs of left heart chambers overload, and risk of endocarditis.[3] The traditional treatment is surgical repair, which was performed for the first time by Lillehei et al. in 1954. The surgical approach is considered to be a gold standard, but it's associated with high morbidity and mortality.[3,10,11] Surgery is generally a safe procedure, but it does have some potential risks, including complete atrioventricular block (cAVB) in 1–5%, significant residual VSD in 1–10%, the necessity for re-operation in 2%, and even death in 0.6–5%. Furthermore, infections, tachy-arrhythmias, and neurological complications may occur. [3,10] In the last decade, percutaneous approaches to the closure of VSD have been developed in order to reduce the impact of such drawback of surgery.[3,10] It has less psychological impact , shorter hospitalization, lower cost, less pain and discomfort, and there is no need for admission to an intensive care unit.[3,10-13] Since the first VSD closed by a transcatheter approach by Lock et al., various devices and techniques have been used, such as the Rashkind double umbrella, the Bard Clamshell, the Amplatzer septal, duct or muscular VSD occluder, or the Gianturco coils.[3,10,14] However, only the recent introduction of Amplatzer muscular VSD occluder and perimembranous VSD occluder has increased the number of subjects in whom percutaneous closure is feasible.[15]

In this case, percutaneous procedure was the choice in managing the cardiac disorder of the patient. Because it had many advantages compare to surgery. Lower risk of developing infection and bleeding, were also supported the consideration.

The Gianturco coil has been used for over three decades with US FDA approval for "vascular occlusions" in humans.[6,16-19] Cesare Gianturco, an Italian radiologist, first described the use of coiled spring "wooly tails" in 1975. Since that time, Gianturco-type coils have become commonplace in catheterization labs for occlusion of a variety of vascular (and other) structures.[16,17] Coils come in a variety of shapes, sizes, and constituents, but share the common characteristic of a spring-like shaped metal coil, usually embedded with synthetic filaments that promote thrombosis . The most common coils in pediatric catheterization labs are made of steel or platinum. Steel coils are "MRI safe," but produce greater artifact than platinum coils.[17]

In 1992, Cambier and Moore reported the successful use of a standard 0.038″ Gianturco occlusion coil for a transcatheter PDA occlusion. Within a very short time thereafter, Gianturco coils became used routinely for the occlusions of the PDA in multiple centers in the US and throughout the rest of the world.[16] The Gianturco coil proved to be effective, very safe and relatively cheap for the occlusion of the small to moderate diameter PDA.[19] Physicians throughout the world (including even the United States) who were caring for patients with a patent ductus soon accepted the Gianturco coil occlusion as the standard treatment and the standard of care for treatment of the PDA.[17,19] Latiff et al (1999) used Gianturco coil to close multiple muscular VSDs in a 10-month old boy. These authors successfully deployed 4 and 3 coils, respectively, for a 3.5-mm apical defect and 1.5-2.0 mm mid-muscular defect. A small residual shunt was found at 3-month follow-up with the patient's clinical feature improved significantly. Thus in special situation, Gianturco coils might be also used to close a muscular VSD.[5,6]

In this case, from the catheterization revealed muscular VSD with the diameter of 2,88 mm. Percutaneous closure procedure was then conducted using Gianturco coil with diameter of 8 mm. The size of 8 mm was chosen based on the size of the defect estuary on the left ventricle, which was 6 mm, added with 2. The procedure was using general anesthesia. It was considered successful. There was no complication appeared during the procedure. Three hours after the coil implantation, the patient was sent back to the ward with stable vital sign.

Complication of percutaneous closure consists of residual shunt, which may spontaneously closed in 3 to 6 months, conduction abnormalities (cAVB), and device embolization. However, in the current era, even if recently published data is not available, the rate

of cAVB is probably at the lowest limit of the range.[3,10] However, compared with surgery in which cAVB usually appears early after the operation, in patients treated percutaneously, the occurrence of cAVB is quite unpredictable and it is usually a late problem. This complication is related to the proximity of the conduction system to the margins of the muscular VSD. 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 are no specific data about the mechanisms involved in the occurrence of cAVB after percutaneous closure of a muscular VSD. Large studies are needed to clarify the real impact of arrhythmic problems in these patients and the mechanism of the events.[10,21]

Our patient was discharged in a good condition 1 day after the coil implantation. Aspirin was continued to be given until 6 months after closure. Before discharged, the patient was conducted echocardiography evaluation and then repeated at 2 weeks after the closure. There was small residual shunt observed, but no pericardial effusion. The patient had no complaint. The patient was planned for follow-up with echocardiography at the 1, 6 and 12 months after the procedure.

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