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BMH Medical Journal 2014;1(3):56-63 **Point of View**

Device Closure of Small Ventricular Septal Defects: When and Why?

IB Vijayalakshmi, MD, DM, DSc.

Sri Jayadeva Institute of Cardiovascular Sciences and Research, Bangalore - 560069, India

Address for Correspondence: Dr. IBVijayalakshmi, MD, DM, DSc, Professor of Pediatric Cardiology, Sri Jayadeva Institute of Cardiovascular Sciences and Research, Bangalore - 560069, India. Email: docvj@yahoo.com

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Introduction

Ventricular septal defect (VSD) accounts for approximately 20-30 % of all forms of congenital heart disease (CHD). They were traditionally closed surgically in the past. The surgery though safe carries the risk of morbidity, complete heart block, wound infection and thoracotomy scar on the chest. On weighing the risk and benefit, the small VSDs were not submitted to surgery in the past. So for many years it has been taught that the small VSDs are to be left alone and surgery is not recommended. Many clinicians believed in spontaneous closure of VSDs. Hence they advised the parents to wait for spontaneous closure till the child is 9 years old. But what if the VSD does not close by 10 years is the question. Are they normal as grown up congenital heart (GUCH). The Jane Somerville GUCH unit showed that spontaneous closure occurred only in 10% between the age of 17 and 45 (mean - 27) years in small VSDs in adults. What is worth noting is about 25% had serious complications: infective endocarditis (11%), progressive aortic regurgitation (5%), age-related symptomatic arrhythmias (8.5%) like atrial fibrillation. [1] This means that asymptomatic small VSDs in childhood is not necessarily benign during adult life. This raises the question, when we have a safe non-surgical device closure available to close the small VSDs, should we put the future lives of the young in danger by not giving the benefit of technology to them?

Background

The advent of Amplatzer Muscular VSD occluder device with higher closure rates and safety of the procedure made it an attractive alternative to the surgical closure, as it avoids the cardiopulmonary bypass. Today other Amplatzer VSD devices have also been designed to close perimembranous VSD and the results have been very encouraging. [2] Now there are reports of Amplatzer duct occluder II (ADO II) specially designed to close long ducts in small infants used successfully in closing small VSDs (<6 mm). [3] Hence, today non-surgical percutaneous transcatheter closure of VSDs has revolutionized the management of VSDs. Now that non-surgical device closure of small VSDs is possible without the hassles of surgery the question is being asked when and why to close the small VSDs. In this review the transcatheter device closure of small VSDs will be discussed with examples and the technical aspects in detail.

When to call VSD a small defect ?

In small VSDs the dimension is less than one third the size of the aortic annulus diameter or less than 0.5 square cm with a significant systolic pressure gradient between the ventricles and the pressure in the right ventricle being normal. They may be isolated or multiple as in Swiss cheese VSDs. The isolated small VSDs have normal cardiac anatomy and function. Since the right ventricular pressure is normal and there is not much increase in pulmonary flow there is no damage to the lung arterioles and the heart functions normally. A systolic thrill and a prominent murmur heard through a stethoscope is usually the only sign that brings the VSD to attention during school health check up. This murmur is commonly noted during the first week of life. Children with such VSDs appear healthy, have no growth retardation or congestive heart failure (CHF). Cardiomegaly is absent. Thus a small defect that has no hemodynamic effect, may produce a thrill and murmur out of proportion to its clinical significance. Chest x-ray: Cardiac contour and size are normal. ECG: It is usually normal.

Nearly one-third to one-half of all small VSDs close spontaneously (on their own). This seemingly miraculous event occurs most often before the baby is 1 year old, almost always before age 4 (75% by 2 years of age). The closure is due to the small VSD being located between heart fibers that increase in size in time, thus encroaching upon the opening in the ventricular septum. Even if a small VSD does not close spontaneously, surgical repair is usually not recommended for small VSDs. So one should understand the natural history, which has a wide spectrum, ranging from spontaneous closure to death due to complications. Also 90% of VSDs that close do so by age 8 years. [4,5] Hence many clinicians advise the parents to wait for the spontaneous closure till 8 years without checking the child at regular intervals. But one should think of what happens to other half of patients who do not have spontaneous closure of VSD, when they become adults and also clearly understand the various methods of spontaneous closure, because it is always at the cost of causing another abnormality. For example: 1. The in growth of fibrous tissue with endocardial proliferation with septal aneurysm (**Figure 1A**), may cause thromboembolism. 2. Adherence of septal tricuspid leaflet resulting in tricuspid regurgitation. 3. Prolapse of aortic cusp through the defect leading to aortic regurgitation (AR). The AR is due to Venturi effect (**Figure 2**), seen more in subpulmonic VSDs (30%) than perimembranous VSDs (5-8%). The occurrence of AR is more between the ages of 5-9 years (**Figure 1B**). 4. Closure of VSD can occur with vegetation of infective endocarditis, but this may lead to death.

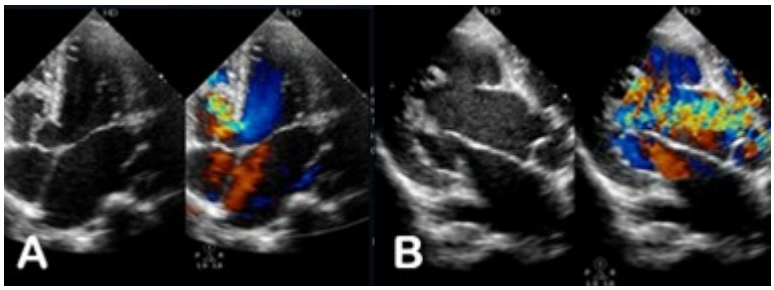

Figure 1 A: Transthoracic echocardiography (TTE) in apical four chamber view shows large subaortic VSD (12 mm) with septal aneurysm with 6mm opening. B: TTE in parasternal long axis view with color Doppler shows small VSD with coronary cusp prolapse and severe AR in 6 years old boy.

So if we understand the natural history of the small VSDs we realize that it can also pose serious problems if child is not carefully monitored and the small VSD is not closed in time. Instead of simple device closure that requires few minutes, it may end up with major surgery with aortic valve replacement!

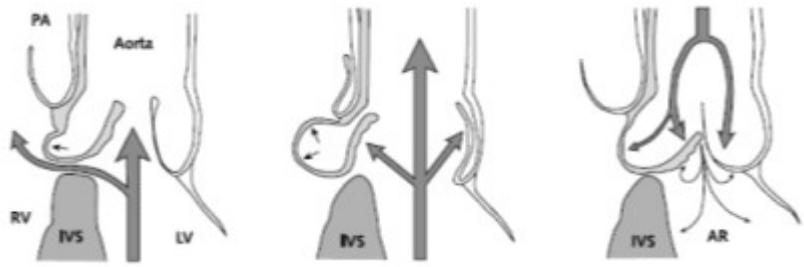

Figure 2: Shows the schematic representation of systole and diastolic movement of coronary cusp causing AR

Case 1

5 years old boy a known case of small VSD weighing 12 Kgs, was treated as bronchopneumonia with parental antibiotics and IV fluids in a local hospital. As the fever continued for 1 ½ months, and his condition deteriorated, he was referred for further management, because he became very moribund and developed CHF. The X-ray chest of the child showed increased bronchovascular markings with non-homogenous shadow in left lower lobe (as compared to his old X-ray - **Figure 3 A** and **B**), which the pediatrician thought to be bronchopneumonia. Probably that shadow was due to pulmonary embolism as the emergency transthoracic echocardiography (TTE) in apical four chamber view showed a large highly mobile vegetation (looking like a bunch of grapes) attached to tricuspid valve (TV) causing tricuspid stenosis (TS), and tricuspid regurgitation (TR) leading to CHF (**Figure 4 A** and **B**). The boy was hospitalized and treated for 6 weeks with high antibiotics after blood culture grew coagulase negative staphylococcus.

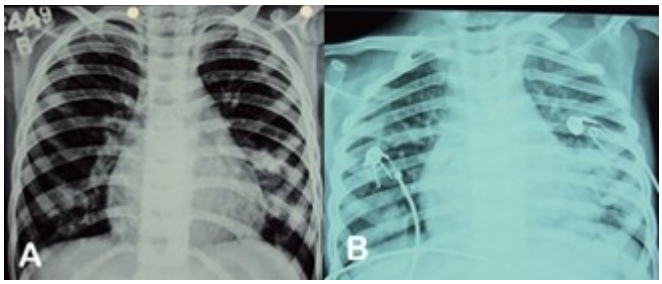

Figure 3 A: Normal X-ray chest in PA view, note no plethora in 5 years old patient with small VSD (old). B: present X-ray showed cardiomegaly, dilated right atrium, increased bronchovascular markings with non-homogenous opacity in left lower lobe.

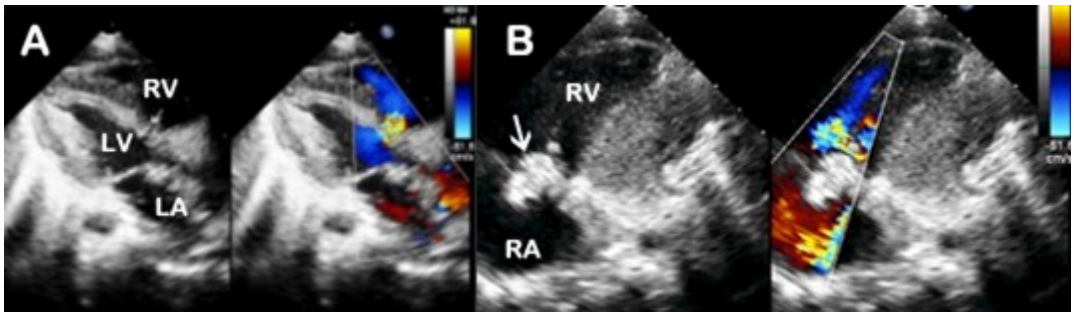

Figure 4: A. Parasternal long axis view with the color Doppler shows a small sub aortic VSD with normal left ventricle (LV) and left atrium (LA). B: Apical four chamber view shows dilated right ventricle (RV) and right atrium (RA) with large vegetations (arrow) on tricuspid valve causing tricuspid stenosis and tricuspid regurgitationin a 5 years old boy diagnosed as small perimembranous VSD in infancy.

This case raises the question, is sub acute bacterial endocarditis prophylaxis enough in India or

should we close the small VSD with the device, so that children do not die of infective endocarditis (IE)?

Case 2

A 2 years old boy known case of small VSD (Type A Gerbode defect) presented with history of fever for one month. TTE showed dilated left ventricle (LV) and right atrium (RA), pericardial effusion (PE) and a large, long highly mobile vegetation attached to the RA end of Gerbode defect with trivial TR (**Figure 5 A** and **B**). The TV was normal with trivial TR indicating TV is not affected by IE. The child was in CHF. Before he could be stabilized and referred for surgery, the child suddenly died due to pulmonary embolism.

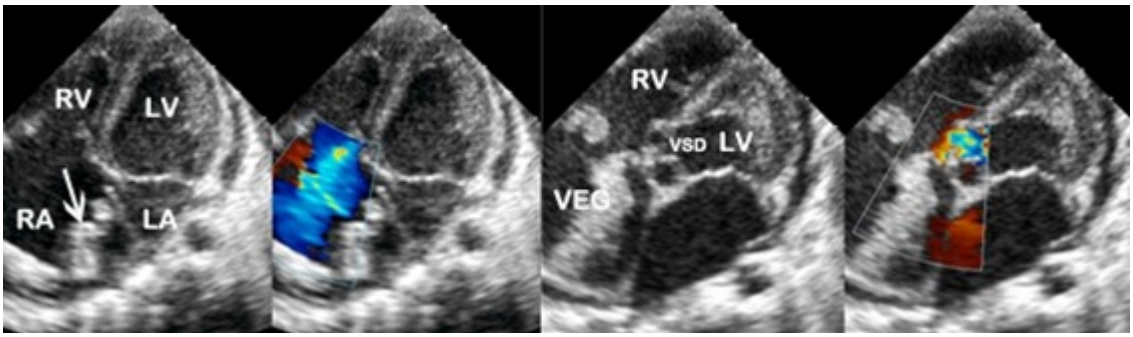

Figure 5: A: TTE in a 2 years old boy shows dilated right atrium (RA) and left ventricle (LV), pericardial effusion (PE) and a large, long vegetation (arrow) attached to the RA end almost closing the Gerbode defect, with normal tricuspid valve and with trivial tricuspid regurgitation on color Doppler. B: Apical five chamber view with color Doppler shows LV to RA, left to right shunt closed by large vegetation (arrow) attached to VSD.

This case raises the question, the Gerbode defects are basically small VSDs that do not close spontaneously. So are we justified in leaving them as small and not closing them with device, when ADO II appears to be tailor made for closing Gerbode defect. If we follow the most recent recommendations of the American Heart Association, unrepaired VSDs don't require endocarditis prophylaxis! So we do not give endocarditis prophylaxis and do not close them by device as the evidence based medicine does not recommend, are we justified in allowing them to die?

Case 3

10 months old boy weighing 6.5 Kgs, presented with repeated lower respiratory tract infection (LRTI) and failure to thrive was diagnosed as Gerbode defect measuring 5 mm on TTE was taken up for device closure. Pulmonary artery pressure was 25/15mean 18 Qp:Qs was 1.8:1. Left ventricular angiogram done in AP view showed the contrast opacifying the RA (**Figure 6A**). Through the right Judkins catheter in the ascending aorta, 0.018 Terumo guide wire was used to cross the defect, over which 4F multipurpose catheter was passed. The Terumo was exchanged with J tipped 0.025 guide wire and was anchored in superior vena cava (Figure 5B). Over the J guide wire 4F Shuttle sheath was passed. Then 5x6 ADO II was deployed so that the distal retention disc was in RA, the middle lobe at the defect and the LV disc in the LV under the echocardiographic guidance. Hand injection of contrast done from side port of Y connector demonstrated the ADO II device in situ (**Figure 6C**) but still attached to screw. The device is released after check echo (**Figure 7**).

There are 2 types of Gerbode defects (**Figure 8**) which can be diagnosed by careful interrogation of TTE in apical four chamber view with the color Doppler. The type A is direct type in which the blood in the LV, shunts through the small area of the membranous septum and this communication is above the TV (**Figure 9A**). The type B is the indirect type which is more common, where the flow of blood is from the LV through a VSD into the RV and then through a defect in the septal leaflet

of TV into the RA. [6] This communication occurs below the TV and many echocardiographers mistake it for TR (**Figure 9B**).

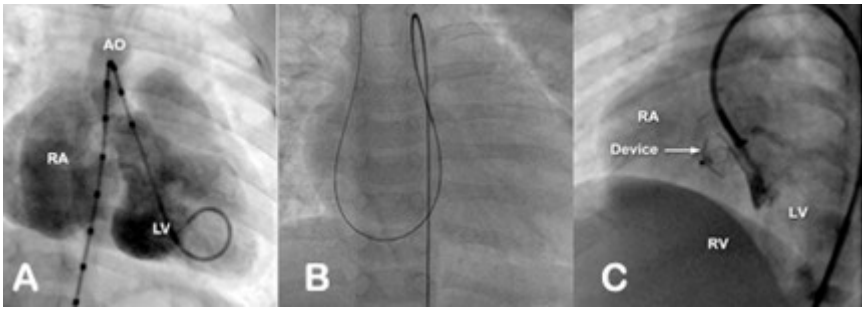

Figure 6 A: Left ventricular (LV) angiogram done in AP view showed the contrast opacifying the dilated right atrium(RA) through the defect B: Illustrates Left heart catheter through the defect on a J tipped 0.025 guide wire anchored in superior vena cava. C:Hand injection of contrast in demonstrates device (arrow) in situ.

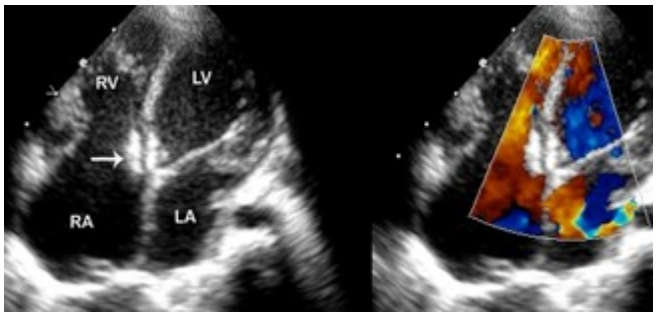

Figure 7: TTE in apical four chamber view with color Doppler demonstrates ADO II device in position with no mitral or tricuspid regurgitation.

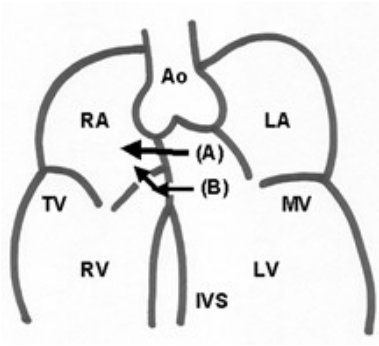

Figure 8: The diagrammatic illustration of type (A) - direct type in which shunt is from LV through the small area of the membranous septum and this communication is above the TV. The type (B)- indirect type the shunt is from the LV through a smallVSD into the RV and then through a defect in the septal leaflet of TV into the RA.

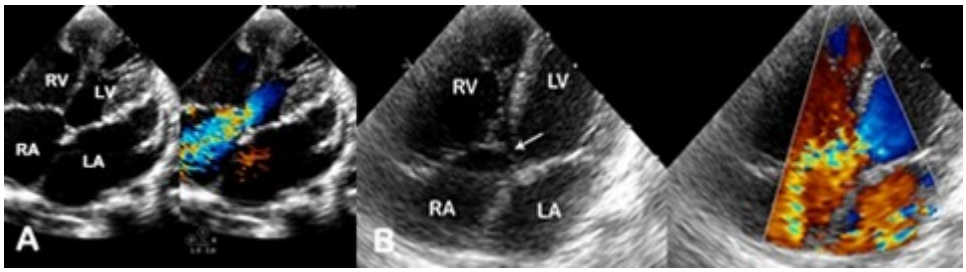

Figure 9: A: Apical four chamber view with color Doppler shows direct defect between LV and RA (Type A) B: Apical four chamber view with color Doppler illustrates indirect defect (Type B) shunting through septal leaflet of TV (arrow)

Gerbode defects are rare (0.08%), congenital LV to RA communications. [7] They are traditionally closed surgically with high incidence of complete heart block. But now congenital Gerbode defects can be closed by ADO II with high success rate and very low complication rate. The soft low profile, easily track able ADO II appears to be tailor made for the Gerbode defect, as the central cylinder fits in the defect without compressing the conducting system and the soft retention discs without polyester material do not impinge on the mitral or tricuspid valve. The success rate is very high (100%) with very low complication rate.

Case 4

15 year old boy with past history of recurrent LRTI since infancy, with repeated hospitalizations, failure to thrive, chest pain, palpitations, since child hood, easy fatigability since 1 year. Patient had pectus carinatum, bilateral Harrison sulcus, prominent precordial pulsations, cardiomegaly, short soft systolic murmur in lower sternal border, P2 was loud and palpable. TTE diagnosed CHD - situs solitus, multiple VSDs (large perimembranous VSD - 18 mm and apical VSDs, 4, 2 mm), bidirectional shunt, pulmonary hypertension (PAH) with pressure of 90/68 m 81 mm Hg. He was operated and surgical valved Gortex patch closure of large VSD (> 80% of interventricular septum) was done. The pulmonary artery pressure (PAP) was systemic and decreased to 65% post cardiopulmonary bypass. The post op echo showed small residual apical VSD (4 mm), PAP of 53 mm of Hg. He was discharged on tablet bosentan and sildenafil. He presented 6 months later, with easy fatigability and palpitations and SaO2 - 96%. The TTE showed residual multiple VSDs - mid muscular VSD (4 mm), 2 apical VSDs - 3-4 mm (**Figure 10**), severe tricuspid regurgitation, with PAH (95 mm of Hg). Though the VSDs were small, as they were multiple, so the total shunt was large. Cath revealed Qp:Qs = 2.6 :1, PAP - 95/68 mean 77 and aortic pressure of 106/74 mean 85 mm of Hg.

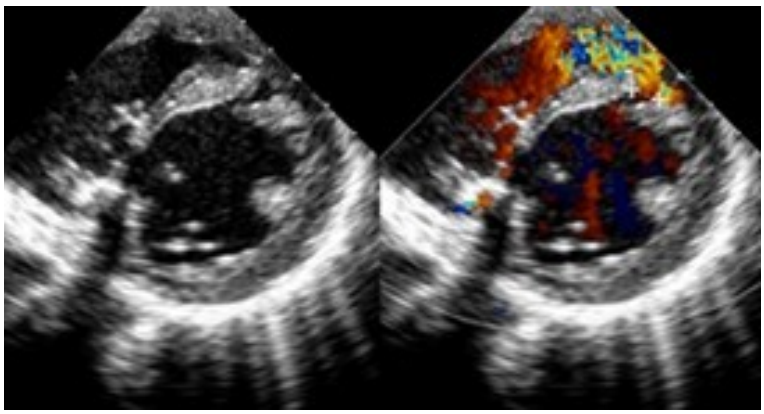

Figure 10: TTE in modified two chamber view with color Doppler shows patch intact, but noncompaction of LV, with multiple Swiss cheese small VSDsin a 15 years old post operative boy.

The LV angiogram in left anterior oblique (LAO) view in this patient showed noncompaction of LV (multiple small arrows indicating deep trabeculae) with two Swiss cheese VSDs (arrows on RV side) with sternal suture wires (**Figure 11A**). Both VSDs were closed by two 6x6 ADO II devices. The check LV angiogram in LAO view shows two ADOII devices (arrows) in situ (**Figure 11 B**). The results of surgery for apical muscular VSDs are often suboptimal owing to difficulties in defect visualization, residual shunting, and ventricular dysfunction.

Closure of the VSD by the transcatheter route is the preferred approach in a select group of patients. Complications with the Amplatzer device are rare in the hands of a skilled operator. Long-term follow-up of device patients is needed.

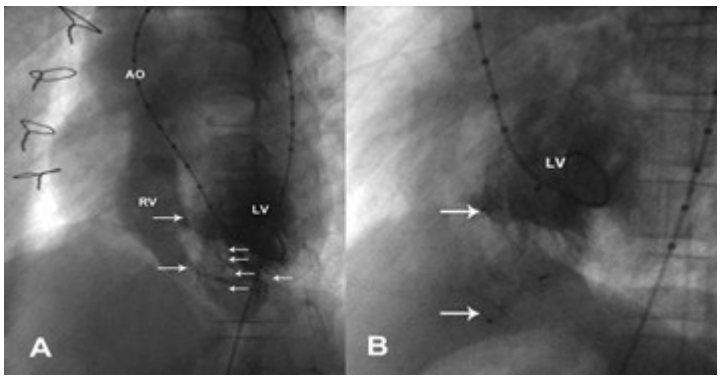

Figure 11: (A) LV angiogram in LAO view in a patient with post-surgical residual VSDs illustrates noncompaction of left ventricle (multiple small arrows indicating deep trabeculae) with two Swiss cheese VSDs (arrows on RV side) with sternal suture wires in 15-year-old boy with pulmonary artery pressure of 90/68 m 81 mm Hg; (B) LV angiogram in LAO view shows two ADO II devices (arrows) in situ with LV noncompaction.

Discussion

The small VSD has been considered benign, not needing any treatment throughout life, other than prophylaxis for a low risk of endocarditis. However, there have been suggestions that this is not always true. [8-10] The reduced left ventricular function on exercise has been reported in adults with small VSD [11] and subnormal working capacity and fractional shortening [12] are reported with small VSDs in GUCH. A term 'ventricularseptal defect cardiomyopathy' [1] was postulated by Bloomfield in 1964. In the Jane Somerville GUCH Unit, [1] one hundred and eighty-eight adults aged 17-72 (mean, 29.2) years with small VSDs were studied. Eighty nine patients (47%) aged 17-44(mean, 26.8) years had no complications through many years, while spontaneous closure occurred in 19 (10%) during adulthood. Forty-six (25%) had serious complications: Twenty-one (11.2%) had IE, AR developed in 37(19.7%) patients; it was severe in nine. The symptomatic atrial arrhythmias (supraventricular tachycardia or atrial fibrillation) occurred in 12 patients (8.5%) and atrial fibrillation was the commonest. In four patients, atrial fibrillation produced severe right-sided congestion with a left ventricular to right atrial shunt and haemodynamic features suggesting 'restrictive cardiomyopathy'. Four patients had ventricular arrhythmia. Disproportionate left ventricular enlargement on echocardiography and/or chest radiography was present in 26 (13.8%) without lesions to account for it. In this group AR, IE and left ventricular abnormalities appeared equally frequent in a separate analysis of the subgroup without associated lesions, whereas arrhythmias and restrictive physiology might be less common in the absence of other lesions. The course of small VSDs appears not entirely benign. The incidence of important complications (IE, severe AR, symptomatic arrhythmia, frank heart failure) was 24.5% over 13 years series. Bacterial endocarditis is still a serious risk to be aware of and atrial arrhythmias become more frequent after the age of 40 years and can cause important deterioration. The major indications for VSD closure is prevention of endocarditis, and AR. Endocarditis is associated with a 10% mortality rate in patients with a congenital VSD. A history of IE is class I indication for closure. [13]

The nonsurgical transcatheter device closure of VSD is promising and exciting. It is revolutionary approach to the treatment of patients with a clinically important VSD. The defects can be closed without a sternotomy, without the need for cardiopulmonary bypass, and with a relatively short recovery time and hospital length of stay. Hence device closure is indicated in small VSDs 1. To prevent IE, AR. 2. In multiple Swiss cheese VSDs. 3. Post-surgical residual VSDs and 4. In rare Gerbode defects. It is imperative that the care we provide should not become evidence-averse.

Conclusion

Device closure in childhood would decrease the incidence of bacterial endocarditis and possibly the

development of AR and arrhythmias. The ADO II is the appropriate device for closure of VSD <6 mm and especially so in infants and smaller children. ADO II is safe, effective and an attractive alternative to surgical closure of Gerbode defects and small perimembranous VSDs with septal aneurysms. It provides a less invasive alternative to open heart surgery/redo surgery. Adverse events like transient junctional rhythm and complete heart block are rare and are easily manageable.

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