ORIGINAL ARTICLE

# Pattern of Ventricular Septal Defects and Associated Complications

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## ABSTRACT

**Objective**: To determine the frequency of various types of Ventricular Septal Defect (VSD) in local population and their complications.

**Study Design**: Case series.

**Place and Duration of Study**: The Paediatric Cardiac Unit of the Children Hospital and Institute of Child Health, Lahore, from January 2006 to December 2007.

**Methodology:** All patients aged between one day to 18 years presenting to the unit with isolated VSD during the study period were evaluated using 2-D, continuous wave Doppler and colour Doppler techniques on transthoracic echocardiography. Where necessary, transesophageal echocardiography and/or angiography data was used to further clarify the anatomy.

**Results**: Of the 854 patients with isolated VSD, 710 were of perimembranous type (83.1%), 100 were muscular type (11.7%), 25 were doubly committed subarterial (DCSA) type (2.9%) and 19 patients were having inlet VSD (2.2%). The mean age was 2.1±3 years. Females were 332 (38.8%) and males were 522 (61.1%). Aortic Valve Prolapse (AVP) was seen in 77 patients (10.8%). The mean age for this subgroup was 5.8±4 years with 54 (70.1%) males and 23 (29.9%) females. Of those, 36 (47.3%) had various degree of aortic regurgitation. Right ventricular outflow tract obstruction of variable severity was found in 4 patients. Two of these patients had echocardiographic evidence of subacute bacterial endocarditis.

**Conclusion**: Perimembranous (PM) VSD is the commonest type of ventricular septal defect presenting to a tertiary care hospital. The incidence of AVP and AR is high (10.8%) and was found almost entirely in perimembranous outlet VSDs rather than DCSA. This observation has special implication in Pakistan where pool of unoperated VSDs is large.

**Key words:** *Ventricular septal defect (VSD). Aortic valve prolapse. Aortic regurgitation.*

## INTRODUCTION

Ventricular Septal Defects (VSDs) result from a deficiency of growth or a failure of alignment or fusion of component parts of ventricular septum.[1] Isolated Ventricular Septal Defect (VSD) is the most common congenital cardiac malformation responsible for 25-45% of all congenital heart diseases.[2] The most popular and practical classification of VSD is that described by Soto *et al.*,[3] dividing VSDs into perimembranous, muscular and doubly committed subarterial (DCSA) types. Perimembranous defects account for 80% of surgical and autopsy series. Muscular VSDs account for 5-20% of the defects and generally have a better prognosis.[4] They tend to close spontaneously earlier than perimembranous VSD.[5] Doubly committed subarterial VSDs account for 5-7% of surgical and autopsy series. These are much more common in Asia, especially in the Far East (about 29%).[6]

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The natural history of VSD is characterized by many complications. Of special interest is prolapse of the aortic valve, which classically occurs with doubly committed subarterial and less commonly with perimembranous outlet type. The reported incidence varies depending upon the type of study, the age group studied and the race. These patients usually have deficiency of muscular and fibrous support below the aortic valve with herniation of the right coronary leaflet through the VSD and development of aortic regurgitation. These patients may have associated infundibular pulmonary stenosis. Less commonly, prolapse of non-coronary cusp may occur in perimembranous VSD. The prolapsed aortic leaflet partially closes a moderate to large VSD and limits the left to right shunt. The prevalence of this complication increases with age but can occur before 6 years of age.[7]

Aortic regurgitation can also occur in these patients and varies in severity in different individuals. The anatomic and hemodynamic features in doubly committed subarterial VSD have a great impact on the development of aortic valve leaflet deformity and subsequent aortic regurgitation.[8] Cusp deformity (imbalance of width) may predict possible progressive deterioration of aortic regurgitation.[9] Aortic regurgitation

is progressive in nature and presence of even mild aortic regurgitation or aortic valve prolapse in the absence of aortic regurgitation is an indication for surgery.

Perimembranous outlet VSDs are also associated with infundibular hypertrophy,[10] and right ventricular outflow tract obstruction can progress in severity. This also requires surgical intervention.

The purpose of the present study was to determine the frequency of various types of Ventricular Septal Defect (VSD) in patients with isolated VSD at a tertiary care paediatric cardiac unit with special reference to complications.

## METHODOLOGY

It was a cross-sectional descriptive echocardiography-based study in paediatric patients referred to a tertiary care cardiac centre. It included all isolated VSD cases, aged between one day and 18 years seen from January 2006 to September 2007, who underwent detailed echocardiography at Paediatric Cardiology Unit of Children Hospital and Institute of Child Health, Lahore.

Accuson Aspen model and GE vivid 3 with color Doppler systems were used for echocardiography. All echocardiographic were performed and/or viewed by paediatric cardiologists experienced in echocardiography for CHD. Transthoracic echocardiography was performed in all cases using 2D, continuous wave Doppler and colour Doppler technique, while transoesophageal echocardiography was used very selectively, if indicated. Patients having isolated Ventricular Septal Defect (VSD) (absence of any other major cardiac anomaly) only were included in the study. Patients having minor associated anomaly, like a small patent ductus arteriosus, a small secundum atrial septal defect and mild mitral regurgitation were also included. VSDs were classified as PM, DCSA, muscular and inlet VSDs using Soto's classification.[3] Records of the selected patients were reviewed to assess the frequency of various types of VSD with special reference to aortic valve prolapse and aortic regurgitation. Severity of aortic regurgitation was assessed by using parameters like left ventricular end – diastolic and systolic dimensions, Doppler flow velocity measurement and assessment of length, width and area of regurgitant jet.[11,12] Findings were graded as mild, moderate and severe according to these parameters. Subpulmonary obstruction or any other complication was also noted. The severity of subpulmonary obstruction was assessed by Doppler peak flow velocity measurement across the RVOT and by using the modified Bernoulli equation.[13] The data thus collected was analysed using Microsoft Excel.

The ethical committee of the hospital approved of the study.

## RESULTS

A total of 7756 patients with congenital and acquired heart disease underwent echocardiography during this period. Eight hundred fifty four had isolated VSDs (11.01%). The mean age was 2.1±3 years. Of those, 83.1% were perimembranous type, 11.70% were muscular type, 2.92% were DCSA type and 2.2% were inlet type (Table I).

A total of 77 cases with aortic valve prolapse were seen, of whom, 76 had Right Coronary Cusp (RCC) prolapse and only one had Non-Coronary cusp (NCC) prolapse. One case had doubly committed subarterial (DCSA) type of VSD, all the other patients had perimembranous outlet type of VSD i.e. out of a total of 710 perimembranous VSD, 76 had aortic cusp prolapse (10.8%). The mean age of those patients was 5.8±4 years with 54 males and 23 females. Eight patients had RVOT obstruction, 6 patients had mild degree of obstruction, while 2 patients had moderate and severe obstruction each (Table II).

Acquired RVOT obstruction was found in 8 out of 729 patients with perimembranous type VSD. It was mild in most cases while only 2 patients with moderate or severe RVOT obstruction were found.

Echographic evidence of subacute bacterial endocarditis was found in 9 children (about 1%). It consisted of vegetations in 7 patients and additional rupture sinus of valsalva in 2 patients. All these patients were older than 2 years and vegetations affected both left and right sides of heart equally.

The percentage of patients with large VSD and severe pulmonary hypertension was 3.1% in this study.

**Table I**: Types of VSDs (n=854).

| Type | Subtype | Number | Percentage |
|---|---|---|---|
| PM VSD | | 710 | 83.1% |
| Muscular VSD | | 100 | 11.7% |
| | Apical | 20 | 20% |
| | Mid | 49 | 49% |
| | Upper | 16 | 16% |
| | Multiple | 9 | 9% |
| | Coexisting with PM | 5 | 5% |
| DCSA | | 25 | 2.9% |
| Inlet | | 19 | 2.2% |
| Total | | 854 | 100% |

**Table II**: Aortic cusp prolapse (n=77).

| Type | Subtype | Number | Percentage |
|---|---|---|---|
| PM VSD | | 710 | |
| | Aortic valve prolapse | 76 | |
| | No AR | 41 | 53.9% |
| | Mild AR | 26 | 34.2% |
| | Moderate AR | 09 | 11.8% |
| | Severe AR | 01 | |
| DCSA | Aortic valve prolapse | 1 | |
| Total | | 77 | |

## DISCUSSION

The natural history of VSD is not only dictated by the size of the VSD but also the type of the defect. Classically DCSA VSD is associated with progressive development of aortic valve prolapse and aortic

regurgitation and incidences reported to be high in Chinese and Far Eastern population.[5] The commonest type was of perimembranous VSD in this study. The second in order of frequency were muscular VSD (11.7% of total VSD) and least frequent were DCSA type, which accounted for 2.9% of the total. These results were more in keeping with what is found in Western literature, where the largest group of VSD consists of PM type, muscular and DCSA following in decreasing order of frequency.[6]

There are very few local studies on this subject. In a study at NICVD, Karachi, Aziz *et al.* found that PM VSD were 92% of total VSD, DCSA were 7% and the least common were muscular i.e. 1.7%.[14] But in this study, the largest group of patients were older than one year (68% of patients). In the present study, however, the ages of patients were between one day and 18 years with mean age of 2.1 years, and muscular VSDs were found mostly in younger patients (74% of the patients were younger than one year). It may be that small muscular VSD tend to close earlier than PM.[6] In another study by Sadiq *et al.,* the incidence of VSD was 32% of all Congenital Heart Diseases (CHD) in patients presenting to a tertiary care paediatrics cardiology unit.[2]

In contrast, in Southeast Asian children the studies done on VSD types showed that DCSA was quite common reaching upto 29-30% of total VSD. Among the SAARC countries, there were very few studies done on the types of congenital heart disease. In one Sri Lankan study, VSD was 27.5% of total congenital heart disease.[15]

Aortic valve prolapse was present almost exclusively in patients with PM outlet type. There was only one case of DCSA with aortic valve prolapse, while 76 patients out of 729 cases of PM VSD had RCC prolapse (10.4% of total PM VSD). This frequency is quite high and is in keeping with other studies. Lue *et al.* found aortic valve prolapse and AR in 11.9% of their patients with VSD.[7] Brauner *et al.* found AV prolapse in over 5% of children with VSD.[16] In yet another study Ando *et al.* found 16% cases of RCC prolapse in patients of VSD.[17]

Aortic valve prolapse was almost exclusively affecting the right cusp. Only one patient was found with prolapse of the non-coronary cusp, while none of the patients had both cusps prolapsing simultaneously. Chiu *et al.* found that in DCSA, the prolpase cusp was always the RCC but in perimembranous outlet VSD besides RCC.[10] NCC involvement was also significant e.g. 16.5%.[10] In another study, Somanath *et al.* found RCC prolapse in 48%, NCC prolapse in 41% and both (RCC and NCC prolapse) in 11% of their patients with perimembranous VSD.[18]

The mean age of the subjects having AV prolapse was 5.8 years. Chiu *et al.* found that the mean age of onset of AV prolapse in their patients with DCSA and PM outlet type of VSD were 4.9 and 5 years respectively.[10] Somanath *et al.* found that the mean age of their subjects with VSD and aortic valve prolapse was 13.4 years. Their study group included adult patients also, with an age range of 2-45 years.[18]

In this study, 47% patients with AV prolapse were having some degree of aortic regurgitation. Majority (72%) had only mild or trivial AR, 28% had moderate AR and only one child had severe AR with LV dysfunction. Somanath *et al.* found grade 1 and 2 AR in 51% of patients and grade 3 and 4 AR in 48%. Their study group consisted of older patients suggesting that severity of AR may progress with increasing age.[8]

Detection of RCC prolapse is critical in patients with outlet VSD because this complication may cause permanent aortic regurgitation.[19] Careful echocardiographic evaluation is needed to identify various combination of progressive RV obstruction or aortic valve prolapse.[20] Patients with clinically important aortic regurgitation or RVOT obstruction are candidates for surgery.[21] Early repair may prevent progression of aortic insufficiency.[22]

Although less frequent, discovery of this RVOT obstruction is clinically important because it may cause RV hypertrophy. Glenn *et al.* found that 5.8% patients of VSD developed infundibular stenosis while AV prolapse was found in 3.6% of their patients.[23] In this study, old RVOT was found in 8 out of 729 patients.

A large VSD is associated with severe pulmonary hypertension and exposes the patient to risk of developing pulmonary vascular disease. This is the major indication of surgery in patients with large VSDs. This aspect was, however, not studied any further.

## CONCLUSION

Perimembranous (PM) VSD was the commonest type of ventricular septal defect studied in this series. The frequency of AVP and AR was high (10.8%) and was found almost entirely in perimembranous outlet defect rather than DCSA. This observation has special implication in Pakistan where pool of unoperated VSDs is large.

## REFERENCES

1. Rhodes LA, Keane JF, Keane JP, Fellows KE, Jonas RA, Castaneda AR. Long follow-up (43 years) of ventricular septal defect with audible aortic regurgitation. *Am J Cardiol* 1990; **66**: 340-5.
2. Sadiq M, Roshan B, Latif F, Bashir I, Sheikh SA, Khan A. Pattern of paediatric heart disease in Pakistan. *J Col Physicians Surg Pak* 2002; **12**:149-53.
3. Soto B, Becker AE, Moulaert AJ, Lie JT, Anderson RH. Classification of ventricular septal defects. *Br Heart J* 1980; **43**:332-43.
4. Hrahshah AS, Hijazi IS. Natural and modified history of ventricular septal defect in infants. *Pak J Med Sci* 2006; **22**:136-40.

5. Miyake T, Shinohara T, Nakamura Y. Fukuda T, Tasato H, Toyohara K, *et al.* Spontaneous closure of ventricular septal defects followed up from < 3 month of age. *Pediatrics Intern* 2000; **46**:135-40.
6. McDaniel NL, Gutgesell HP. Ventricular septal defect. In: Allen HD, Driscoll DJ, Sheddy RE, Feltes TF (edi). Moss and Adams' heart disease in infants, children and adolescent. 7th ed. Philadelphia: *Lippincott Williams & Willkins*; 2008.p.667-82.
7. Lue HC, Sung TC, Hou SH, Wu MH, Cheng SJ, Chu SH, *et al.* Ventricular septal defect in Chinese with aortic valve prolapse and aortic regurgitation. *Heart Vessels* 1981; **2**:111-6.
8. Kobayashi J, Koike K, Senzaki H, Kobayashi T, Trunemoto M, Ishizawa A, *et al.* Correlation of anatomic and hemodynamic features wtih aortic valve leaflet deformity in doubly committed subarterial ventricular septal defect. *Heart Vessels* 1999; **14**:240-5.
9. Tomita H, Arakaki Y, Ono Y, Yamada O, Yagihara T, Echigo S. Imbalance of cusp width and aortic regurgitation associated with aortic cusp prolapse in ventricular septal defect. *Jpn Circ J* 2001; **65**:500-4.
10. Chiu SN, Wang JK, Lin MT, Wu ET, Lu FL, Chang CI, *et al.* Aortic valve prolapse associated with outlet-type ventricular septal defect. *Ann Thorac Surg* 2005; **79**:1366-71.
11. Teague SM, Heinsimer JA, Anderson JL, Sublett K, Olson EG, Volyes WF, *et al.* Quantification of aortic regurgitation utilizing continuous wave doppler ultrasound. *J Am Col Cardiol* 1986; **8**: 592-9.
12. Perry GJ, Helmeke F, Nanda NC, Byard C, Soto B. Evaluation of aortic insufficiency by doppler color flow mapping. *J Am Col Cardiol* 1987; **9**:952-9.
13. Lima CO, Shan DJ, Valdes-Cruz LM, Goldberg SJ, Barron JV, Allen HD, *et al.* Non-invasive prediction of transvalvular pressure gradient in patients with pulmonary stenosis by quantitative two-dimensional echocardiographic doppler studies. *Circulation* 1983; **67**:866-71.
14. Aziz K. Ventricular septal defects. In: Heart disease in children. 2nd ed. Karachi: *The Authors;* 2000.p.254-77.
15. Wickramasinghe P, Lamabadusuriya SP, Narenthiran S. Prospective study of congenital heart disease in children. *Ceylon Med J* 2001; **46**:96-8.
16. Brauner R, Birk E, Blieden L, Sahar G, Vidne BA. Surgical management of ventricular septal defect with aortic valve prolapse: clinical considerations and results. *Eur J Cardiothoracic Surg* 1995; **9**:315-9.
17. Ando M, Takao A. Pathological anatomy of ventricular septal defect associated with aortic valve prolapse and regurgitation. *Heart Vessels* 1986; **2**:117-26.
18. Somanath HS, Gupta SK, Reddy KN, Murthy JS, Rao AS, Abraham KA. Ventricular septal defect with aortic regurgitation: a hemodynamic and angiographic profile in Indian subjects. *Indian Heart J* 1990; **42**:113-6.
19. Hori Y, Yoshimura N, Kimura M. Right coronary cusp prolapse, evaluation by MRI. RSNA presentation Dec 2004 Hall D, Lakeside Centre.
20. Wu MH, Wang JK, Chang CI, Chill IS, Leu HC. Implications of anterior septal malignment in isolated ventricular septal defect. *Br Heart J* 1995; **74**:180-5.
21. Webb GD. Challenges in the care of adult patients with congenital heart defects. *Heart* 2003; **89**:465-9.
22. Yetkin G, Refile B. An unusual case of chrysomonas luteala in Infective endocarditis in a patient with ventricular septal defect. *Indus Universatisi tip facultesi Dergisi* 2005; **12**:193-5.
23. Glen S, Burns J, Bloomfield P. Prevalence and development of additional cardiac abnormalities in 1448 patients with congenital ventricular septal defects. *Heart* 2004; **90**:1321-5.