# Ventricular Septal defect and associated complications

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## Abstract

**Objective:** To determine the frequency of various types of ventricular septal defects (VSD) and associated complications in local paediatric population.

**Methods:** A cross sectional descriptive study was conducted on children undergoing echocardiography in a single centre from January 2006 to December 2009 at Paediatric Cardiology Department, Ch. Pervaiz Elahi Institute of Cardiology Multan- Tertiary referral centre for paediatric and adult cardiac services in South Punjab.

The data on all children below 15 years of age undergoing detailed transthoracic two-dimensional echo and doppler studies was reviewed. Cases with isolated ventricular septal defects were studied for age of presentation, gender, type, and associated complications. The data was analyzed with SPSS 11 version.

**Results:** A total of 5018 patients with congenital heart diseases underwent echocardiography during this period. A total of 1276 patients had isolated VSD (25%). Mean age was 3.1 ± 3.64 years (range: 1 day to 15 years). Females were 440 (34.5%) and males were 836 (66.5%). Of 1276 patients, 1014 (79.3%) were Perimembranous type, 124 (9.8%) were muscular type, 85(6.7%) were doubly committed subarterial type and 53 (4.2%) inlet VSD. Small, moderate and large VSDs were 428(33.6%), 443(34.7%) and 405(31.7%) respectively. Severe pulmonary hypertension was noted in 286 (22.4%) cases. Aortic valve prolapse was present in 85 (6.7%) cases and varying degrees of aortic valve regurgitation was seen in 67 (5.2 %) patients.

Right ventricular outflow tract obstruction was found in 21 (1.6%) cases. Left ventricular outflow tract obstruction was noted in 09 (0.7%) cases. Echo evidence of infective endocarditis was present in 06 (0.5 %) patients.

**Conclusion:** Perimembranous ventricular septal defect was found to be the commonest type of ventricular septal defect. Large ventricular septal defects usually lead to severe pulmonary hypertension. Severe pulmonary hypertension was the commonest complication followed by Aortic Valve Prolapse and Aortic Regurgitation. Rest of the complications were rare.

**Keywords:** Venticular septal defect, Aortic cusp prolapse, Aortic regurgitation, Severe pulmonary hypertension (JPMA 61:1001; 2011).

## Introduction

Ventricular septal defect (VSD) is a developmental defect of the interventricular septum resulting from a deficiency of growth or a failure of alignment or fusion of component parts of ventricular septum.[1] Isolated ventricular septal defect occurs in approximately 2-6 of every 1000 live births and accounts for more than 15-20% of all congenital heart diseases.[2]

Soto et al divided VSD into Perimembranous, Muscular and doubly committed subarterial (DCSA) types.[3] Perimembranous defects are the most common types of ventricular septal defects and account for 80% of such defects.[4] Perimembranous ventricular septal defects are associated with pouches or aneurysms of the septal leaflet of the tricuspid valve, which can partially or completely close the defect.[5] Muscular VSD account for 5-20% of the defects and generally have a better prognosis. They tend to close spontaneously earlier than perimembranous VSD.[6] Doubly committed subarterial VSD account for 5-7% of surgical and autopsy series. These are much more common in Asia, especially in the Far East (about 29%).[7]

The natural history has a wide spectrum, ranging from spontaneous closure to congestive heart failure (CHF) to death in early infancy.[8] Spontaneous closure frequently occurs in children, usually by age of 2 years. Closure is most frequently observed in muscular defects (80%), followed by perimembranous defects (35-40%). Outlet ventricular septal defects have a low incidence of spontaneous closure, and inlet ventricular septal defects do not close.[5]

The natural history of VSD is also characterized by many complications. Of special interest is prolapse of the aortic valve cusp, which classically occurs with doubly committed subarterial and less commonly with perimembranous outlet type.[9] Secondary aortic insufficiency, is associated with prolapse of aortic valve cusps. This complication is observed only in 5% of patients with ventricular septal defect.[10] Aortic regurgitation is due to a poorly supported right coronary cusp combined with the Venturi effect produced by the ventricular septal defect jet, resulting in cusp prolapse.[11] 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.[12]

Perimembranous outlet VSD are also associated with infundibular hypertrophy, and right ventricular outflow tract obstruction can progress in severity. This also requires surgical intervention.[13]

Discrete fibrous subaortic stenosis is occasionally associated with a ventricular septal defect. This complication is most often reported with perimembranous ventricular septal defects and can first appear after either spontaneous or surgical closure.[14] Infective endocarditis is rare in children younger than 2 years.[15]

Pattern of VSD and associated complications is already known in the literature but we wanted to highlight our own pattern of this defect in South Punjab as few studies are available from this area.

## Patients and Methods

This was a cross-sectional descriptive echocardiography based study, conducted in department of Pediatric Cardiology, Ch. Pervaiz Elahi Institute of Cardiology Multan, a tertiary referral centre for cardiac patients in South Punjab. Study was conducted from January 2006 to December 2009. A total of 1276 patients were included by consecutive sampling. All new children below fifteen years of age with suspected acyanotic congenital heart disease referred to a single tertiary referral centre were analyzed. The diagnosis was primarily made on echocardiography. Size, number and exact location of the defect as well as magnitude of shunt were identified by two dimensional and Doppler echocardiography. Pulmonary artery pressure was estimated by using modified Bernoulli equation. Aortic valve prolapse and aortic regurgitation was also noted. 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.[16] All echocardiograms were performed by two trained paediatric cardiologists using latest GE VIVID-7 DIMENSION echo-machine. Patients with VSD as a part of other congenital cardiac anomalies were excluded from the study.

Data collection procedure: All echocardiography reports were reviewed from hospital record. Patients having isolated Ventricular Septal Defect (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. VSD were classified as Perimembranous, Doubly committed subarterial, Muscular and Inlet VSD using Soto's classification.[3] Functionally VSD was divided into small, moderate and large groups.

Small VSD was defined as a doppler CW gradient across VSD > 60mmHg, no LV dilation and absence of severe pulmonary hypertension.

Moderate VSD was defined as doppler CW gradient across VSD 30-60 mmHg and LV dilation was the absence of severe pulmonary hypertension.

Large VSD was a doppler CW gradient across VSD < 30mmHg LV dilation may or may not be present. Presence of severe pulmonary hypertension.

Records of the selected patients were reviewed to assess the frequency of various types of VSD. Associated complications like severe pulmonary hypertension, aortic valve prolapse and aortic regurgitation, acquired right and left

ventricular outflow tract obstruction and infective endocarditis were also noted. The severity of obstruction was assessed by Doppler peak flow velocity measurement across the right and left ventricular outflow tract and by using the modified Bernoulli equation.[17] The data was reviewed for age of presentation, sex, type, size of VSD and associated complications. Data was entered and processed using SPSS version 11.

## Results

A total of 5018 patients with congenital heart diseases underwent echocardiography during this period. Of these 1276 patients had isolated VSD (25%). Mean age was 3.1±3.64 years (range: 1 day to 15 years). Patients below 2 years were 62%, when they came to seek medical evaluation. Females were 440 (34.5%) and males were 836 (66.5%). Male to female ratio was 2:1.

Patients were classified according to Soto's classification[3] as Perimembranous, Muscular, Doubly committed subarterial type and Inlet VSD. Distribution of patients with different types of VSD is presented in Table-1. Of 1276 patients, 1014 (79.3%) were Perimembranous type, 124 (9.8%) were Muscular type, 85(6.7%) were Doubly committed subarterial type and 53 (4.2%) were having Inlet VSD. According to functional classification there were 428(33.6%) small, 443(34.7%) moderate and 405(31.7%) large size defects.

**Table-1: Types of VSD (n=1276).**

| Type | Number | Percentage |
|---|---|---|
| Perimembranous | 1014 | 79.3 |
| Muscular | 124 | 9.8 |
| DCSA | 85 | 6.7 |
| Inlet | 53 | 4.2 |

**Table-2: Complications associated with VSD.**

| Complications | Number | Percentage (%) |
|---|---|---|
| Severe pulmonary hypertension | 286 | 22.4 |
| Aortic cusp prolapse | 85 | 6.7 |
| Aortic regurgitation | 67 | 5.2 |
| RV outflow tract Obstruction | 21 | 1.6 |
| LV outflow tract Obstruction | 09 | 0.7 |
| Infective endocarditis | 06 | 0.5 |

**Table-3: Aortic cusp prolapse and aortic regurgitation.**

| Subtype | Complication | Percentage (%) |
|---|---|---|
| | **Aortic cusp prolapse** | |
| Perimembranous (1014) | 73 | 7.1 |
| Doubly committed subarterial VSD(85) | 12 | 14 |
| | **Aortic regurgitation** | |
| Perimembranous (1014) | 58 | 5.7 |
| Doubly committed subarterial VSD(85) | 09 | 10.5 |

Complications were noted in 37.1% (474) of total cases. Severe pulmonary hypertension was the most common complication associated with large VSD and it was noted in 286 (22.4%) patients. Severe pulmonary hypertension was associated with LV dilation in 230 cases (81%) and most of the patients in this group were below two years.

The most common complication seen with small and moderate VSD was aortic cusp prolapse and aortic regurgitation followed by right and left ventricular outflow tract obstruction and then infective endocarditis. About, 85 (6.7%) cases were having right aortic cusp prolapse and varying degrees of aortic valve regurgitation was seen in 67 (5.2 %) patients. This complication was observed more frequently with perimembranous type of VSD than reported in literature. While, 73 (85%) cases belonged to perimembranous type. Out of these 58 (86%) were associated with Aortic valve regurgitation and 12 (14%) patients with DCSA VSD contributed to aortic cusp prolapse and 9 had aortic regurgitation. (Table-3) Acquired right ventricular outflow tract obstruction was found in 21 (1.6%) cases. Left ventricular outflow tract obstruction was noted in 09 (0.7%) cases and 06 (0.5%) patients had Echo evidence of infective endocarditis.

## Discussion

Ch. Pervaiz Elahi Institute of Cardiology Multan is the only cardiac centre in Southern Punjab catering to a population of almost 30 million. Catchment area extends to parts of Sind and Balochistan. Isolated Ventricular Septal Defect was found in 25% of patients with congenital heart diseases analyzed during the study period. Similar study in Children hospital and institute of child health, Lahore conducted in 2006- 2007 showed 11% incidence of VSD.[18] Yet another study at NWFP showed similar results.[19]

The commonest type was of perimembranous VSD in our study. The second in order of frequency were muscular VSD (9.8%) and least frequent were Doubly committed subarterial type, which accounted for 6.7% of the total. These results were more in keeping with what is found in Western literature, where the largest group of VSD consists of perimembranous type, muscular and doubly committed subarterial type in decreasing order of frequency.[7] There are very few local studies on this subject. In a study at NICVD, Karachi, Aziz et al. found that perimembranous VSD were 92% of total VSD, doubly committed subarterial type were 7% and the least common were muscular i.e. 1.7%.[20] However, in this study, the largest group of patients were older than one year (68% of patients) and the ages of patients were between one day and 15 years with mean age of 3.1±3.64 years, and muscular VSD was found mostly in younger patients. It may be that small muscular VSD tend to close earlier than perimembranous.[7] Similar results were shown in local studies by Uzma Kazmi et al and Masood Sadiq et al

conducted in Lahore.[18,21] Aortic valve prolapse was present in 6.7% of total patients. This frequency is in keeping with other studies. Lue et al.[9] found aortic cusp prolapse and aortic regurgitation in 11.9% of their patients with VSD. Brauner et al. found aortic cusp prolapse in over 5% of children with VSD.[22] In yet another study Ando et al[23] found 16% cases of right coronary cusp prolapse in patients of VSD.

Classically Doubly committed subarterial type VSD is associated with progressive development of aortic cusp prolapse and aortic regurgitation. Contrary to this, our study showed that incidence of aortic cusp prolapse and aortic regurgitation with perimembranous outlet VSD was higher than previously noted in literature. About, 7.1% of perimembranous outlet VSD had aortic cusp prolapse. This finding is in accordance with a study conducted at Lahore, where 10.4% of perimembranous outlet VSD was having prolapsed aortic cusp.[18]

Aortic regurgitation was seen in 86% perimembranous VSD and 14% doubly committed subarterial VSD. Similar findings were noted by Somanath et al.[24] They found 84% perimembranous and 16% doubly committed subarterial VSD with aortic regurgitation.

Glenn et al[25] found that 5.8% patients of VSD developed infundibular stenosis. In the present study right ventricular outflow tract obstruction was found in 1.6% of cases.

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 VSD. Severe pulmonary hypertension was noted in 22.4% cases and it was seen almost exclusively with a large VSD.

Limitations of this study are that it does not give the incidence or prevalence of ventricular septal defect and its complications in total population as it was confined to one hospital attendance. Also, excluded were children not reaching a tertiary care centre due to poor access to medical facilities, yet, results are comparable with other local and international studies. As it was a retrospective study, it was difficult to control bias and confounders. Also, we had to rely on the available written record. Results are, at best, hypothesis-generating.

## Conclusion

Perimembranous ventricular septal defect was found to be the commonest type of ventricular septal defect. Severe pulmonary hypertension was the commonest complication seen with large ventricular septal defects. Aortic cusp prolapse and aortic regurgitation was commonly found with small and moderate ventricular septal defect.

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