Original Article

Study of Types and Complications of Venticular Septal Defect in Our Institute

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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 May 2013 to January 2016 at CVTS Department Grant Medical College ,Mumbai, Maharashtra.

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.

Results: A total of 70 patients with congenital heart diseases underwent echocardiography and surgical procedure during this period. A total of 15 patients had isolated VSD (21.4%). Mean age was 3.1 ± 3.64 years (range: 4 years to 15 years). Females were 5 (33.3%) and males were 10 (66.6%). Of 15 patients, 11 (73.3%) were Perimembranous type, 2 (13.3%) were muscular type, 1(6.66%) were doubly committed subarterial type and 1 (6.66%) inlet VSD. Small, moderate and large VSDs were 5(33.3%), 6(40 %) and 4(26.6 %) respectively. Severe pulmonary hypertension was noted in 5 (33.3%) cases. Aortic valve prolapse was present in 5 (33.3%) cases and varying degrees of aortic valve regurgitation was seen in 3 (20 %) patients.

Right ventricular outflow tract obstruction was found in 1 (6.66%) case. No Echo evidence of infective endocarditis.

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.

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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] [Figure 1]

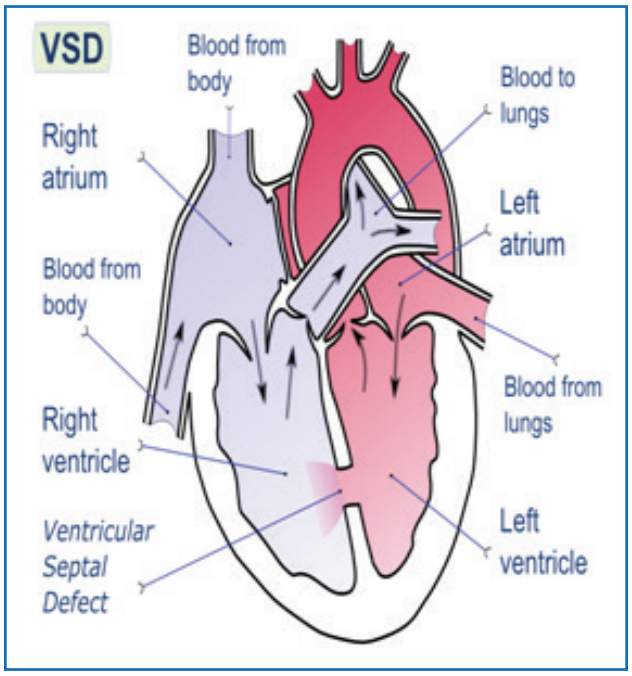

Fig. 1: Ventricular Septal Defect [Source-Sabiston textbook of cardiac surgery]

Soto et al divided VSD into Perimembranous, Muscular and doubly committed subarterial (DCSA) types.[3] [Figure 2] 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. 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%).[6]

Ventricular septal defect is usually symptomless at birth. It usually manifests a few weeks after birth.VSD is an acyanotic congenital heart defect, aka a left-to-right shunt, so there are no signs of cyanosis in the early stage. However,

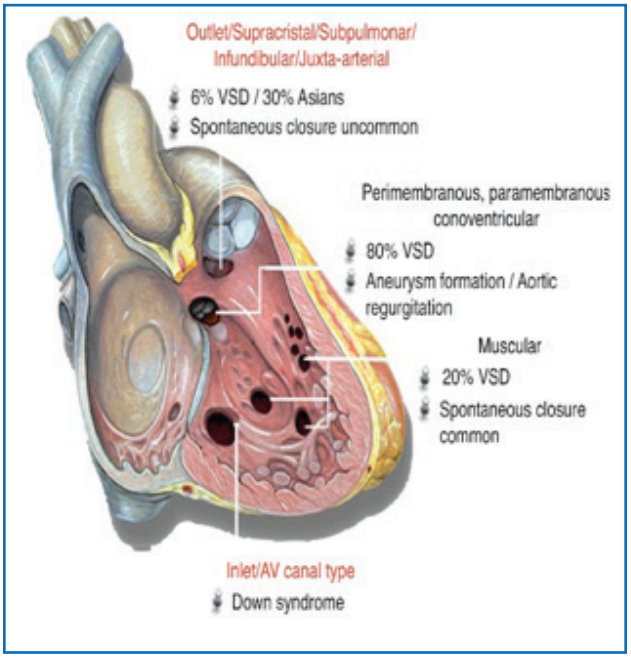

Fig. 2: Types of Ventricular Septal Defect [Source-Sabiston textbook of cardiac surgery]

uncorrected VSD can increase pulmonary resistance leading to the reversal of the shunt and corresponding cyanosis.

Pansystolic (Holosystolic) murmur along lower left sternal border (depending upon the size of the defect) +/- palpable thrill (palpable turbulence of blood flow). Heart sounds are normal. Larger VSDs may cause a parasternal heave, a displaced apex beat (the palpable heartbeat moves laterally over time, as the heart enlarges). An infant with a large VSD will fail to thrive and become sweaty and tachypnoeic (breathe faster) with feeds.

The restrictive VSDs (smaller defects) are associated with a louder murmur and more palpable thrill (grade IV murmur). Larger defects may eventually be associated with pulmonary hypertension due to the increased blood flow. Over time this may lead to an Eisenmenger's syndrome the original VSD operating with a left-to-right shunt, now becomes a right-to-left shunt because of the increased pressures in the pulmonary vascular bed

The natural history has a wide spectrum, ranging from spontaneous closure to congestive heart failure (CHF) to death in early infancy. 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.[7] Secondary aortic insufficiency, is associated with prolapse of aortic valve cusps. This complication is observed only in 5% of patients with ventricular septal defect.[8] 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.[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.[10] Perimembranous outlet VSD are also associated with infundibular hypertrophy, and right ventricular outflow tract obstruction can progress in severity. This also requires surgical intervention.[11] 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.[12] Infective endocarditis is rare in children younger than 2 years.[13] Pattern of VSD and associated complications is already known in the literature but we wanted to highlight our own pattern of this defect in our area.

Materials and Methods

This was a cross-sectional descriptive echocardiography based study, conducted in department ofCVTS,Grant Medical College,Mumbai,Maharashtra. Study was conducted from May 2013 to January 2016. A total of 70 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. 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 ductusarteriosus, 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.[14]The data was reviewed for age of presentation, sex, type, size of VSD and associated complications.

Result

A total of 70 patients with congenital heart diseases underwent echocardiography during this period. Of these 15 patients had isolated VSD (21.4%). Mean age was 3.1±3.64 years (range: 4 years to 15 years). Females were 5 (33.3%) and males were 10 (66.6%). Male to female ratio was 2:1.

Patients were classified according to Soto's classification as Perimembranous, Muscular, Doubly committed subarterial type and Inlet VSD. Distribution of patients with different types of VSD is presented in Table-1.

Table 1: Types of VSD [n-15]

Type	Number	Percentage[%]
Perimembranous	11	73.3
Muscular	2	13.3
DCSA	1	6.66
Inlet	1	6.66

Of 15 patients, 11 (73.3%) were Perimembranous type, 2 (13.3%) were Muscular type, 1(6.66%) were Doubly committed subarterial type and 1 (6.66%) were having Inlet VSD. According to functional

classification there were 5(33.3%) small, 6 (40 %) moderate and 4(26.6 %) large size defects. Complications were noted in 37.1% (15) of total cases. Severe pulmonary hypertension was the most common complication associated with large VSD and it was noted in 5 (33.3%) patients. Severe pulmonary hypertension was associated with LV dilation in 3 cases (60 %) and most of the patients in this group were below eight years. The most common complication seen with small and moderate VSD was aortic cusp prolapse and aortic regurgitation followed by right ventricular outflow tract obstruction . About, 5 (33.3%) cases were having right aortic cusp prolapse and varying degrees of aortic valve regurgitation was seen in 3 (20 %) patients. This complication was observed more frequently with perimembranous type of VSD than reported in literature. Acquired right ventricular outflow tract obstruction was found in 1 (6.66 %) cases. Left ventricular outflow tract obstructionand infective endocarditis was not noted in any patients.

Table 2: Complications associated with VSD

Complications	Number	Percentage[%]
Severe pulmonary hypertension	5	33.3
Aortic cusp prolapse	5	33.3
Aortic valve regurgitation	3	20
RV outflow tract obstruction	1	6.66
Infective endocarditis	0	0

Discussion

The commonest type was of perimembranous VSD in our study. The second in order of frequency were muscular VSD (13.3%) and least frequent were Doubly committed subarterial type, which accounted for 6.66% 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. However, in this study, the largest group of patients were older than five year (68% of patients) and the ages of patients were between 4 years to 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.[1] Aortic valve prolapse was present in 33.3 % of total patients. This frequency is in keeping with other studies. Lueet 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.[15] In yet another study Ando et al[16] 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. Glenn et al[17] found that 5.8% patients of VSD developed infundibular stenosis. In the present study right ventricular outflow tract obstruction was found in 6.66% 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 33.3% 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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Competing Interests

Not Declared

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