Original Article

The Pattern of Ventricular Septal Defects and the Severity of Associated Pulmonary Hypertension in Our Set-Up

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ABSTRACT

Background*:* Ventricular Septal Defect (VSD) is commonest of all the congenital heart diseases. This is found as an isolated lesion as well as in association with other congenital cardiac lesions. The management and outcome of isolated VSD is not only dependent upon the size but also depends on the associated complications of ventricular septal defect (VSD). Pulmonary hypertension is not only the most common complication but also the most important indication of surgery in our setup.

Objectives*:* To study the pattern of various types of ventricular septal defects (VSDs) and assessment of severity of associated pulmonary hypertension in our population.

Study Design*:* Cross sectional descriptive study.

Patients and Methods: The study was conducted at the paediatric cardiology department of The Children Hospital & The Institute of Child Health Multan, from October 2009 to March 2011. All patients with ages between 1 day to 15 years presenting with isolated VSD during the study period were studied using 2-D, continuous wave Doppler and color Doppler transthoracic echocardiography.

Results: Out of 403 patients with isolated VSD, 288 were of perimembranous type (71.4%), 57 were of muscular type (14.2%), 19 were of doubly committed sub arterial (DCSA) type (4.7%) and 39 patients were having inlet VSD (9.7%). The mean age was 2.4 years. Females were 137 (34.0%) and males were 266 (66.0%). Pulmonary hypertension was present in 210 patients (52.1%). Amongst these mild pulmonary hypertension was present in 86 (40.9%), moderate in 65 (30.9%) and severe pulmonary hypertension was present in 59 (28.1%).

Conclusion: Perimembranous (PM) VSD is the commonest type of ventricular septal defect presenting to our hospital. The incidence of pulmonary hypertension is very high (52.1%) and even severe pulmonary hypertension was found in about a quarter of the patients. This shows the degree of delay in surgery and the major reason is non availability of pediatric cardiac surgery centers in government setups.

Key Words: Ventricular Septal Defect (VSD), pulmonary hypertension,

INTRODUCTION

Ventricular septal defect (VSD) is the most common congenital cardiac malformation[1]. VSD can be classified in many ways but the most popular and practical classification is dividing VSD's into perimembranous, muscular and doubly committed sub arterial (DCSA). It was described by Soto et al[2]. Perimembranous defects account for 80% of surgical and autopsy series. Muscular VSD's account for 5 – 20% of the defects and generally have a better prognosis[3]. They tend to close spontaneously earlier than perimembranous VSD[4]. Doubly committed sub arterial VSD's account for 5 – 7% of surgical and autopsy series but they are much more common in Asia, especially in Far East where incidence is about 29% [5].

Un operated VSD is associated with many complications. These include failure to thrive, repeated chest infections, development of pulmonary hypertension, prolapse of aortic valve cusps along with aortic regurgitation, right or left ventricular outflow tract obstruction, pulmonary vascular obstructive disease and even Eisenmenger syndrome. Pulmonary hypertension is of special consideration in our setup because it will develop in almost all the cases of moderate to large sized VSDs if left un operated. Furthermore, if it is severe then it may lead to pulmonary vascular obstructive disease and Eisenmenger syndrome and then the patient will be declared inoperable. Prolapse of aortic valve cusp is another important complication. It occurs mainly with doubly committed sub arterial and less commonly with perimembranous outlet type VSDs. The reported incidence varies between 0.7–5percent depending upon the type of age group studied. The prevalence of this complication increases with age but can occur before 6 year of age[6]. Aortic regurgitation can occur in these patients and varies in severity in different individuals. The anatomic and hemodynamic features in doubly committed sub arterial VSD have a great impact on the development of aortic valve leaflet deformity and subsequent aortic regurgitation[7]. Cusp deformity may

predict possible progressive deterioration of aortic regurgitation[8].

Perimembranous outlet VSDs are also associated with infundibular hypertrophy and right ventricular outflow tract obstruction can progress in severity and may lead to right ventricular hypertrophy[9].

The purpose of the present study was to identify the relative incidence of type of ventricular septal defect (VSD) in patients with isolated VSD and to assess the severity of associated pulmonary hypertension.

PATIENTS AND METHODS

This is a cross-sectional descriptive study conducted at pediatric cardiology department of Children Hospital Multan from October 2009 to March 2011. It included all isolated VSD cases seen during this period of one and a half years.

Inclusion Criteria:

A) All the patients with ages between 1 day and 15 years.
B) All the patients with isolated VSDs

Exclusion Criteria:

A) Patients with associated congenital cardiac defects other than VSD.
B) Post Op patients with pulmonary artery banding or VSD closure and residual shunt.

Toshiba Pnemio XG machine was used for echocardiography. All echocardiographic studies were performed by pediatric cardiologist experienced in echocardiography for congenital heart diseases. Transthoracic echocardiography was performed in all cases using 2D, continuous wave Doppler and color Doppler techniques. VSDs were classified as perimembranous, doubly committed sub arterial, muscular & inlet VSDs using Soto's classification[2]. Records of these selected patients were reviewed to assess the relative incidence of various types of VSD with special reference to pulmonary hypertension. Pulmonary hypertension was assessed by using CW and color Doppler by measuring tricuspid regurgitation and pulmonary regurgitation jets and by calculating the right ventricular pressure. Systemic blood pressure (LV pressure) was also taken at the same time. Severity was graded in three categories as mild moderate and severe.

Mild PH= RV pressure in between 1/3rd to 1/2 of systemic pressure

Moderate PH=RV pressure in between 1/2 to 2/3rd of systemic pressure

Severe PH= RV pressure more than 2/3rd of systemic pressure

Severity of aortic regurgitation was assessed by using parameters like left ventricular end–diastolic & systolic dimensions, Doppler flow velocity measurement[10] and assessment of length, width and area of regurgitant jet[11]. Sub pulmonary obstruction or any other complication was also noted. The severity of sub pulmonary obstruction was assessed by Doppler peak flow velocity measurement across right ventricular outflow tract[12].

RESULTS

1. Types of VSDs:

Out of 3754 patients who underwent echocardiography during the study period, 403 had isolated VSDs (10.7%). Females were 137 (34%) and males were 266 (66%). Of these 71.4% were perimembranous type, 14.2% were muscular type, 4.7% were DCSA type and 9.7% were inlet type.

Table No. I: Types of VSDs n = 403

Type	Subtype	Number	Percentage
PM VSD		288	71.4%
Muscular VSD		57	14.2%
	Apical	18	31%
	Mid	24	42%
	Upper	15	26%
DCSA		19	4.7%
Inlet		39	9.7%
Total		403	100%

Pulmonary Hypertension in VSD:

A total of 210 (52.7%) cases with pulmonary hypertension were seen. It was mainly associated with larger size defects. The mean age of these patients was 1.8 years. 101 were females and 109 were males.

Table No.2: Pulmonary Hypertension n = 210

Severity	Number	Percentage
Mild	86	40.9%
Moderate	65	30.9%
Severe	59	29.1%

DISCUSSION

VSD is the most common congenital cardiac malformation. Its fate not only depends upon the size of the VSD but also on its type. Larger VSDs tend to have more complications like failure to thrive, repeated chest infections, and development of pulmonary hypertension. Similarly DCSA VSDs and PM VSDs have less chances of spontaneous closure than the muscular VSDs. As regards the overall pattern of types of VSD's, the commonest type was of perimembranous VSD (71.4% cases of total VSD). The second in order of frequency were muscular VSD (14.2% of total VSD) and least frequent were DCSA type, which accounted for 4.7% of the total. These results were similar to those what is found in Western literature, where the largest group of VSD consists of PM type, followed by

muscular and DCSA type in decreasing order of frequency[6].

Locally very little work has been done on this aspect of the disease. 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%[10]. Whereas, in Southeast Asian children the studies done on VSD types showed that DCSA was quite common reaching up to 29 to 30% of total VSDs[7]. Among the South Asian countries, there were very few studies done on the incidence of the types of congenital heart disease. In one study, done in Sri Lanka VSD was 27.5% of total congenital heart diseases[11]. In another study done in Lahore, Pakistan by Sadiq M. et al the incidence of VSD was 32% of all congenital heart diseases[12].

In our study, 52% of the patients had some degree of pulmonary hypertension. Amongst these 52% patients, 40.9% had mild, 30.9% had moderate and 28.1% had severe pulmonary hypertension. While overall incidence of severe pulmonary hypertension was found to be 14.6%. This is a very high percentage as compared to western studies where incidence of severe pulmonary hypertension in VSD cases is less than 5%[13]. Moreover 52% incidence of overall pulmonary hypertension is again a very high and alarming figure as compared to the studies done in South East Asia. In our study, incidence of pulmonary hypertension was very high even among the patients of less than I year of age. 11% of the patients presenting under I year of age had severe pulmonary hypertension while this incidence is found to be very low in study done by Levi DS et al[14].

To detect aortic valve prolapse is critical in patients with PM and DCSA VSDs because this complication may cause permanent aortic regurgitation[15]. In our study, 4.2% of the patients had aortic valve prolapse. Patients with clinically important aortic regurgitation or RVOT obstruction are candidates for surgery[16]. Early repair may prevent progression of aortic valve damage and regurgitation. Right ventricular outflow tract obstruction was found in 3 out of 288 patients with perimembranous type VSD of our study. Glenn et al found that 5.8% patients of VSD developed infundibular stenosis while AV prolapse was found in 3.6% of their patients[17].

All this shows that pulmonary hypertension is the most alarming complication of VSD in our setup. This is because of lack of freely available pediatric cardiac consultation facilities in Pakistan. Moreover delay in surgery even after the diagnosis and proper medical management is another important factor. This shows that how much it is necessary to develop new pediatric cardiac surgery centers in this country.

CONCLUSION

Amongst the types of VSDs, perimembranous VSD is the most common type and DCSA VSD is the least common type. The incidence of pulmonary hypertension is very high. This signifies that to develop new pediatric cardiac surgery centers in Pakistan is the need of the day. We need this because to have early VSD closure is the best way to avoid life threatening complication of pulmonary hypertension.

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