

A Morphological Classification of Ventricular Septal Defects

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ABSTRACT:

BACKGROUND:

The ventricular septal defects are the most common congenital heart disease, Ventricular septum composed of two parts: the membranous septum is relatively small very thin structure and the muscular septum, which is divided into three regions; inlet; trabecular and outlet portion.

OBJECTIVE:

To find out morphological classification of ventricular septal defects in Iraqi population.

PATIENT AND METHOD :

Two hundred isolated ventricular septal defect in 196 patients, were identified by two dimension echocardiography and Doppler color flow mapping.

Age, Sex of patients and family history were studied. Cross section scan was obtained using a SSH-140A Toshiba machine.

RESULTS:

Ventricular septal defects were perimembranous in 70%, inlet in 18%, Trabecular in 9%, and outlet in 3%. Large defects were identified in 21%, and Eisenmenger complex in 5%.

CONCLUSION:

Delay in referral of patient with ventricular septal defects to our hospital usually resulted in delay institution of appropriate management and increase risk of future surgery.

KEY WORDS : ventricular, septal, defect, aortic, prolapse

INTRODUCTION:

The VSD are the most common gross morphological congenital malformation of the heart, accounting for approximately 20% of all cases of congenital heart diseases^(1,2,3).

Defects involving the membranous septum with extension into adjacent inlet, outlet, or muscular septum are termed perimembranous defects (synonymous infracristal, membranous, subaortic)^(1,4).

The inlet defect; are posterior and inferior to the membranous septum, beneath septal leaflet of tricuspid valve.

The muscular defects are classified according to location, apical defects are most common and difficult to visualize from right ventricle, mid muscular defects; which is posterior to trabecula septomarginalis, marginal or anterior defects near the septal-free wall margins^(1,5).

Trabecular defects can occur in combination with other muscular and non muscular defects producing a swiss cheese appearance in the septum.

Finally, the supracristal (infundibular, conal, subpulmonic, or doubly committed subarterial defects) are located in the outflow tract of the right ventricle beneath the pulmonary valve^(1,6,7).

Prolapsing of one of the aortic valve cusps may

occur with outlet or perimembranous VSDs. Patients with outlet defects usually have deficiency of muscular support below the aortic valve with herniation of the right coronary leaflet. The aortic commissure usually are normal. In contrast, patients with perimembranous VSDs have herniation of the right or much less commonly the non coronary cusp, have frequent abnormalities of aortic commissures^(1,8,9,10).

The tendency for VSDs, especially perimembranous and trabecular defects to decrease in size and complete spontaneous closure. Incidence is estimated at 75% to 80% of restrictive defects.

Moderately restrictive and non restrictive defects also spontaneously closed with incidence estimated at 5% to 10%^(2,11,12).

VSDs fall into four anatomic physiologic categories:^(1,2,13)

Small restrictive defects: size of defect less than 1/3 of aortic root, normal pulmonary artery pressure.

1-Moderate restrictive defects: the diameter is less than or equal to that of aortic orifice, and pulmonary artery pressure higher than normal but less than systemic.

2-Large non restrictive defects: the defects size approximately the size of aortic orifice, with identical right ventricle, left ventricle, aorta, and pulmonary artery pressure.

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3-Eisenmenger syndrome: non restrictive VSD with suprasystemic pulmonary vascular resistant and reversed shunt.

AIM OF THIS STUDY:

1-To find out morphological classification of ventricular septal defects in Iraqi population.

2-To determine the prevalence of aortic valve prolapse.

PATIENTS AND METHODS:

Between January 2005 and August 2005 , two hundred isolated ventricular defects in 196 patients, were identified by two dimensioned echocardiograph and doppler color flow mapping . All patients were referred to Ibn-Albitar Hospital for cardiac surgery for evaluation of murmurs from another Hospital or Clinics.

Age of patients at time of diagnosis as having congenital heart disease, age at time of referral, sex of patients, and family history were studied.

Cross-sectional scan were obtained using a SSH-140 A Toshiba machin.

The system are phased-array sector scanners with 2.5, 5, and 7 MHZ frequency transducers.

Estimation of pulmonary artery pressure was done by calculation of trans- VSD pressure drop (gradient).

RESULT:

Table(1) shows the age group of patients .Their age ranged between 1day-18years, the high incidence rate of diagnosis occurs in age group 4wk-2mo. , where as at time of referral the higher incidence rate occurs in age group 2mo.-1years .

There was slight preponderance of female patients with both perimembranous and muscular VSDs, whereas inlet defect were slight more common in males. Family history of VSDs are found in 6.6% of 1st degree relatives of patients, As show on in table(2).

Defect size was classified in 40.5% as small, in 38.5% as moderate, and in 21% as large defects as shown in table(3).

One hundred forty 70% of VSDs , subaortic in location, 18% were inlet in location, 9% trabecular, and 3% subpulmonic defects as shown in figure(1).

Aortic valve prolapse was detected in 10.7% and only one patient with prolapse aortic valve had subpulmonic defect, aortic regurgitation was detected in 62.5% of patients with aortic prolapse as shown in table (6).

Table(7) shows the severity of pulmonary hypertension in study sample. Normal pulmonary artery pressure was detected in all patients with small size defects, and only 5% had Eisenmengers complex.

Table 1: Age groups of patient at time of diagnosis and at time of referral

Age group	Age at diagnosis NO.	%	Age at referral NO.	%
<4 Wks	14	7.14	5	2.55
>4wk-2mo.	100	51	38	19.38
2mo.-1yr.	68	34.69	70	35.71
1yr-5yr.	10	5.1	44	22.44
5yr.-10yr.	4	2	27	13.77
10yr.-18yr.	-	-	12	6.12
Total	196	100%	196	100%

- Mean age of diagnosis 4.5+_{3.4}months.
- Mean age of referral 10.3+_{5.4}months.

Table 2: The distribution of type of VSDs by gender

Type of VSD	Male NO.	%	Female NO.	%	Total	%
Perimembranous	68	34	72	36	140	70
Inlet	19	9.5	17	8.5	36	18
Trabecular	8	4	10	5	18	9
Outlet	3	1.5	3	1.5	6	3
Total	98	49	102	51	200	100

- female : male ratio =1.04:1

Table 3: The distribution of the study sample by the defects

Size of defect	NO.	%
Small	81	40.5
Moderate	77	38.5
Large	42	21
Total	200	100

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Table 4: The relative frequency of different types of VSDs

Type of VSD	NO.	%
Perimemberanous	140	70%
Inlet	36	18%
Trabecular	18	9%
Outlet	6	3%

Table 5: Distribution of aortic valve prolapse by age groups and gender

Age group	Male NO.	%	Female NO.	%	Total	%
<3 years	1	6.25	-	-	1	6.25
3-5 years	2	12.2	1	6.25	3	18.75
5-10 years	6	37.5	4	25	10	62.5
>10	1	6.25	1	6.25	2	12.5
Total	10	62.5	6	37.5	10	100

- Male:female ratio = 1.6:1

Table 6: The distribution of study sample by severity of pulmonary hypertension

Severity of pulmonary hypertension Type of VSDs	Normal	Mild to moderate	Severe	Eisenmengers	Total NO. of patients
Perimemberan-ous	58	55	22	5	140
%	41.5	39.8	15.7	3.5	100
Inlet	15	14	4	3	36
%	41.66	38.88	11.11	8.33	100
Trabecular	8	6	3	1	18
%	44.44	33.33	16.66	5.55	100
Outlet	-	2	3	1	6
%	-	33.34	50	16.67	100
Total	81	77	32	10	200
%	40.5	38.5	16	5	100

DISCUSSION:

In early studies, the perimemberanous type accounted for approximately 80% of defects at surgery or at autopsy, the incidence of muscular VSD being 5 to 20% in same series.^(1,2)

With the advent of echocardiographic technique, this spectrum has changed.

Recently, Trowitzch et al reported echocardiographic finding in 264 consecutive infants aged <1 years and found 68% muscular, 25% perimemberanous, 7% malalignment and 0.7% subpulmonic vsds⁽¹⁴⁾.

Another study from caucasian and black population, confirm that incidence of muscular vsd was 51% and incidence of perimemberanous vsd was 49%⁽¹⁵⁾. we observed a 70% incidence of perimemberanous and 18% incidence of inlet vsd.

Another study also in caucasian and black population confirm that isolated subpulmonic vsd is rare, although it is incidence in japanese and chinese has been reported to be higher (about 30%)^(6,11,15).

The higher incidence of trabecular vsd in infants age less than 2 months, and decrease it is incidence in study samples that deals with different age

groups probably due to high rate of spontaneous closure of muscular VSD by one year of age is almost 2 to 3 times that of perimemberanous VSD, and approximately 90% of muscular VSD close spontaneously within 1 to 10 months of age^(2,9).

The perimemberanous type is the most common (more than 2/3 of the cases), the next in frequency is different in different populations, muscular defects in caucasian and black population^(15,16), outlet defect in japanese and chinese 30%⁽⁶⁾, and inlet defect in iraqi population 18%, probably due to genetic predisposition. A multifactorial etiology has been proposed in which interaction between hereditary predisposition and environment as a cause can not be rule out.

In this study VSDs are slightly more common in female (female 51%, male 49%) and this is nearly similar to what was reported by Hoffman JLE et al.⁽¹²⁾

Anderson C.E. et al was reported positive family history in 3.3% of cases^(2,17). In our series VSDs were founded in 6.6% of 1st degree relative of

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patients who are index cases. probably due to chemical materials that resultwd from the wars in the last two decades.

In this study the diagnosis usually occurs at age of 1to3 months, this result is nearly similar to two studies, one was reported by Samuel B. et al 1988 and the other was reported by vanden heuvel et al 1955^(13,18). This is explained by the fact that the classic harsh holosystolic left sternal border murmur of VSD often does not present until 4 to 6 weeks of age , At this time the normal spontaneous decrease in pulmonary resistance occurs, Which allows for the left ventricular-right ventricular pressure difference to cause ascultatory turbulence of VSD flow^(1,18)

In our study, We observed delay in referring patients to hospital that provide appropriate management to patients with cogenital heart disease. This can explained by family neglection, inadequate and inappropriate explain and education of family about the natural history of disease and risk of developing Eisenmengers complex.

Aortic valve prolapse was deteced in 8% of our patients, aortic regurgitation was detected in 10 (62.5%) of these patients, this result is nearly similar to what was reported by Eroglu, A-G et al , but higher incidence of AVP with subpulmonic VSD 30% was reported by Anzai et al from japan 1991, probably due to high incidence of subpulmonic VSD in Japanese^(6,10).

In our study, defects were classified in 40.5% as small , In 38.5% as moderate, and in 21% as large. In comparison with two studies, one was reported by Eroglu-A-G et al , and other one was reported by Turner et al ,these studies showed higher incidence of small defects 76% and 75% and lower incidence of large defects 6% and 8% respectively.^(19,20)

The true incidence of small VSDs is probably higher than our estimation because of :- 1- some patients with small defect do not seek medical advised, because they are asymptomatic.

2-Two thirds of small defects close spontaneously by one year of age,thus some infant may have had spontaneous closure of VSD in the 1st weeks or months of life before cardiologic evaluation.

Normal pulmonary pressure was recorded in all patients with small defects, severe PA hypertension was recorded in patients with large defect. The primary anatomic variable that determine the pulmonary hypertension of the patients is defect size, it is not related to type of defect .This is explained by the fact that in small or medium sized defects, the size of defect limited the left to right shunt; however, in large defects, there is essentially

no resistance to flow across VSD with systemic pressure in both ventricles and PA.⁽²¹⁾

CONCLUSION:

In Iraqi population the perimemberanous VSD is most common type ,whereas inlet VSD are next in frequency.

Because the type and number of VSDs affect the clinical outcome, precise diagnosis is important. Doppler color flow mapping is an important adjunct to a complete segmental echocardiographic study in identification of VSDs. It increase the sensitivity of the study and can often identify multiple lesion whereas standard non invasive imaging may detect only a single defect.

Aortic valve prolapse rarely occurs before age of 3 years, and aortic regurgitation progress from mild to severe with advancing age.

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