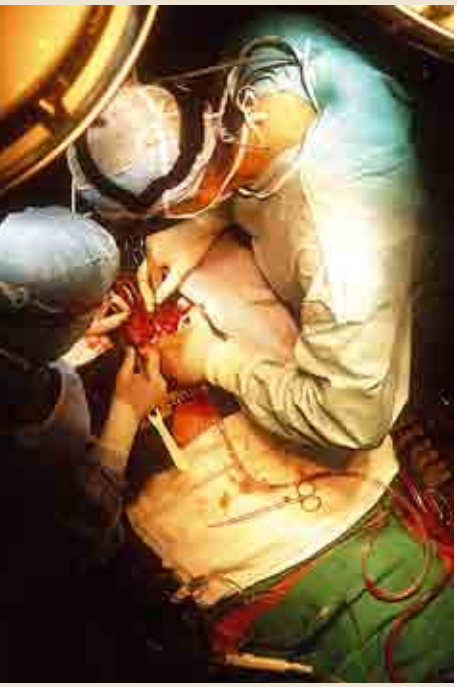
Clinical Course of Ventricular Septal Defect in Children Referred to Aliasghar Center of Zahedan during 2001-2011



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Abstract:

Objective: The aim of this study is a review of clinical progress of ventricular septal defect in children referred to Aliasghar Center of Zahedan during 2001 to 2011.

Method: In this research we have studied all files existing in the archive of the patients referred to Aliasghar Diseases Center of Zahedan, from 2001 to September 2011. The cases with ventricular septal defect diagnosed by echocardiography were selected and required data including location and size of VSD, patient's age along with the manner of VSD closure (spontaneously, surgery, intervention) were collected.

Results: Among 1750 patients with congenital heart diseases, 621 cases (35.5%) were suffering from VSD. In 32.3% of cases, the defect was closed spontaneously, 12.7% underwent closed heart operation, and 53% received medical therapy. Based on the defect location, there were 11.9% muscular, 72.8% perimembranous, 8.1% outlet, and 7.2% inlet type. Also the size of the defect in patients with isolated VSD was as follows: 45.1% small type, 25.9% moderate type, 29% large type. In general, serious complications have been occurred in 4.34% of the patients in long term follow up.

Conclusions: In this study, it has been indicated that the incidence of spontaneously closure of small defects has been increased with time and a large number of moderate defects convert to minor defects by time and few number of large defects spontaneously closed by time. The incidence of serious complications in long term was 4.34% and mostly the results were similar to the results of other studies in this regard.

Key words: Children, Congenital Heart Disease, Ventricular Septal Defect

Introduction:

Congenital heart diseases have been reported in 4-50 thousands of live births. Congenital defects have wide spectrum from severity points of view in children. About 2-3 infants of every 1000 births have symptoms of heart diseases in the first year of their lives. By progress in treatment methods, number of children suffering from congenital heart diseases who survived until adulthood period, have been increased. Despite the latest progress, congenital heart diseases are the first reason of death in patients with congenital anomalies.

Ventricular Septal Defect (VSD) alone

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forms 30-35 percent of the whole heart congenital diseases. VSD also is seen along with other heart diseases including: Tetralogy of Fallot (TOF), complete atrioventricular septal defects, transposition of great arteries (2). Clinical symptoms and natural development of VSD in children are different depending on the defects. The clinical presentation of VSD with small size is mostly a holosystolic murmur and is often without symptoms and in most cases spontaneously closure occur (3).

In newborns suffer from VSD with moderate size; the clinical symptoms are mostly in form of heart failure and growth anomalies. The sign of VSD with moderate size is in form of holosystolic murmur with or without trill in left sternal border and prolonged S2. Also diastolic murmur in heart apex can be osculated in these children. Usually VSDs with moderate sizes have regressed in size by time and even a number of them may be closed spontaneously. Newborns suffering from VSD with large size usually have symptoms during the first month after birth (2) and in two studies on premature and term newborns the incidence of VSD has been reported as 5 % (3).

In the study of Miyake and colleagues study, in order to detect the spontaneously closure of VSD, physical examination, heart catheterization, echocardiography with colored pulse and Doppler were performed. In this study physical examination for long follow up and colored Doppler for short period were performed. In long term reports, the follow up period (10-year), the spontaneously closure of small VSD, was reported as about 75%, however it is reported in other studies as 60%. In short time follow ups (12-month) done by colored Doppler and Doppler pulse, the incidence of spontaneously closure in type of Perimembranous was nearly 45%, and in muscular type it was71.8% (4).

Allen and colleagues reported spontaneously closure during the first 2 years of life between 75-80% in small size and in case of VSDs with moderate sizes, they believed that 15-20% of them may need surgical operation and in case of large VSDs spontaneously closure was occurred in 8 % (5). Most studies have been conducted in patients with VSD along with good results after operation as well as children with normal growth and activities. Long term follow up implicates occurrence of unusual pulmonary hypertension (4%), sinus dysfunction (4%) and aortic valve insufficiency (16%) in VSD with large sizes (6). In another study it was proposed that patients with clinical course and with small to moderate VSD must be followed up in longer period of medical therapy, unless they are affected by heart failure or pulmonary hypertension (7).

Regarding the outbreak and importance of VSD, and since the studies regarding natural course of VSD are limited in our country, we decided to study the 10-year prognosis of VSD in children suffering from this congenital disease in Aliasghar Center of Special Diseases of Zahedan.

Methods:

The study was a case descriptive one and was conducted on all children with VSD referred to Aliasghar Center of Special Diseases of Zahedan from 2001 to 2011. Patients with other anomalies except for VSD were excluded from the study. Regarding the number of referring patients during ten years, the number of subject patients in the study was determined as 621 cases. The sampling method is easy and accessible. The research was conducted by enumeration of the subject society, meaning patients with VSD, in order to reach the desired sample group. The approval of the study protocol was granted and the files of all patients existing in the archive of Aliasghar Center of Special Diseases of Zahedan, from 2001 to September 2011 were studied. Among them, the cases with VSD were divided based on echocardiography and the information existing in the file of every patient, including location and size of the VSD, patient's age along with the manner of closure (spontaneously, surgery, intervention) and the side effects were extracted and entered in the prepared questionnaire form. The patients with bicuspid aorta, mitral valve prolapse and those with patent duct us arteriosus in premature newborns were excluded from the study. Meanwhile the incomplete files were left aside. The data were analyzed using SPSS, descriptive statistical methods of central indexes, dispersing, drawing the table of distribution of plentitude, and drawing chart.

Results:

In this study 621 children with the mean age of 36.2±40.3 (figure -1) and VSD who referred to Aliasghar Center of Special Diseases of Zahedan for diagnosis and treatment during 2001 to 2011 were studied. In this study, 425 children (48.4%) were under 36 months and 196 (31.6%) were over 36 months (table-1) ($p<0.001$). In this study, 339 children of subject patients were boys (54.4%) and 284 (45.6%) were

girls. Among the patients suffering from isolated VSD, in 200 cases (32.2%) the defect was spontaneously closed and in 78 cases (12.6%), the patients required surgical operation for closure of the defect, and 329 cases (53.0%) of them underwent medical therapy (table -2). From among these patients (isolated VSD), based on the defect location, 74 cases (11.9%) had muscular defect, 452 cases (72.8%) had Perimembranous defect, 50 cases (8.1%) were outlet type and 45 cases (7.2%) were inlet type (table -3) ($p<0.001$). The size of defect in spontaneously closure of VSD is an important criterion that among patients with automatic closure of VSD, there were 280 (45.1%) cases with small type, 161 cases (25.9%) with moderate type and 180 cases (29%) with large type, (table -4) ($p<0.001$). Significant side effects in long period of VSD were 4.34%, in general. Among patients with VSD who did not undergo surgical operation, 14 cases affected by Eisenmenger, 3 cases suffered from aorta valve insufficiency that underwent open heart surgery for closure of VSD and surgical repair of aortic valve. Endocardotis was occurred in 3 cases that underwent closed heart operation for closure of VSD. They all recovered after treatment. Also 2 cases that underwent surgical operation for probable Eisenmenger, had no complaints during their follow up that after 10 years from surgical operation. 22 cases (28.2%) had small residual VSD after operation that did not require surgical intervention and 2 cases (2.6%) required further surgical operation and in 1 case, stricture in sub pulmonic valve was occurred that resulted into operation after angiography.

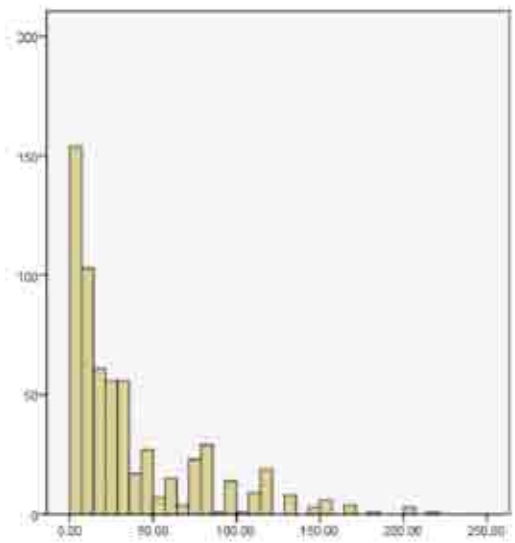

Figure -1: The distribution of patients based on their age

Table -1: VSD Status Based on Age in Patients

Age Group / **Closure**	Under 36 months	Over 36 months	Total	P-value
Spontaneously	110(55.0)	90(45.0)	200(32.2)	
Medicine	272(82.6)	57(17.3)	329(53.0)	
Surgery	43(55.1)	35(44.9)	78(12.7)	P<0.001
Eisenmenger	0(0)	14(100)	14(2.1)	
Total	425(68.4)	196(31.6)	621(100)	

Table -2: Determination of VSD Status Based on Type of Intervention in Patients

Type Of Intervention / Defect Location	Number	Percent
Spontaneously	200	32.2
Medicine	329	53.0
Surgery	78	12.6
Eisenmenger	14	2.2
Total	621	100

Table – 3: Determination of VSD Status Based on Defect Location in Patients

Intervention Type / Defect Location	Spontaneously	Medicine	Surgery	Eisenmenger	Total	p-value
Muscular	43(21.5)	26(7.9)	5(6.4)	0(0)	74 (11.9)	P<0.001
Peri membranous	140(70)	247(75.1)	57(73.1)	8(57.1)	452 (72.8)	
Outlet	11(5.5)	28(8.5)	9(11.5)	2(14.3)	50 (8.1)	
Inlet	6(3)	28(8.5)	7(9)	4(28.6)	45 (7.2)	
Total	200(32.2)	329(53)	78(12.6)	14(2.3)	621 (100)	

Table – 4: Determination of VSD Status Based on Defect Location in Patients

Intervention Type / Defect Location	Spontaneously	Medicine	Surgery	Eisenmenger	Total	p-value
Small	179(89.5)	101(30.7)	0(0)	0(0)	280 (45.1)	P<0.001
Moderate	17(8.5)	143(43.5)	1(1.3)	0(0)	161 (25.9)	
Large	4(2)	85(25.8)	77(98.7)	14(100)	180 (29)	
Total	200(32.2)	329(53)	78(12.6)	14(2.3)	621 (100)	

Discussion:

Among 1750 patients suffering from congenital heart diseases, nearly 35.5% (621 cases) were suffering from Isolated VSD similar to another study that this event was 30-35% (2).

Patients under medical therapy in this center were more than other centers (53%) that this was due to non-acceptance of surgical operation by a large number of them or lack of financial capability of the patients to pay the expenses of heart surgical operation. Among patients in need of surgical operation, 2.2% affected by Eisenmenger due to lack of on time referring (8).

In this research the defect location in 72.8% were Membranous type, 11.9 % muscular type, and 8.1% outlet type, and 7.2% inlet type. In our study, spontaneously closure of muscular VSD was 58%, membranous defect 31%, outlet 22% and inlet 13.3%. The cases resulted in surgical operation were 6.8% in muscular type, membranous 12.6%, outlet 18% and inlet 15.6%. It is worth mentioning that the remaining cases were under medical therapy and follow up due to non-acceptance of surgery by a large number of patients or lack of financial capability (8). Miyake has reported spontaneously closure in muscular type 71.8% and in membranous type 45%, similar to the present search (4 and 9).

In our research, spontaneously closure was 63.9% in small VSD, 10.6% in moderate VSD, and 0.6% in large VSD. Also among patients with spontaneously closure of VSD, 89.5% were small type, however among patients with VSD who underwent surgical operation; in 98.7% the defect was large. In another study spontaneously closure in small type of VSD was 88.6% during the first year of their life (10 and 11). In other study conducted on 882 cases with isolated VSD during 1971-1988, it was indicated that in 22.5% of the patients heart anomaly was due to VSD and in 77.5% of the cases there were no prognosis signs, in 40.2%, VSD was spontaneously closed, in 0.7% there were aortic insufficiency, especially sub pulmonic type of VSD and in 0.5%

bacterial endocarditis have occurred. But nearly 15.5% of VSD cases required open heart operation. The findings of this study were, to large extend, similar to our research (4, 12).

In another study Onat and colleagues concluded that in children with VSD, before puberty period, this defect is also affected. These researchers have also reported that the incidence of spontaneously closure of the defect in their patients who had no increase in pulmonary hypertension about 32%. This research is also coordinated with our research in this regard (13).

In other research, Tuner has studied the clinical course of VSD based on the size of the defect, its location and patient's age and indicated that from 68 subject patients with isolated VSD, in 49 cases (72%) the size of VSD was small, in 14 cases (20.5%) the size of VSD was moderate, and in 5 cases (7.3%) the size of VSD was large. And 35 cases had spontaneous closure and 13 case required surgical operation. Also the researchers of this study showed that from 35 cases of spontaneous closure of VSD (77%), 27 cases were muscular types that this point is also similar to the present research (9,8 and 14).

In the study of Miyake and colleagues study is also shown that the spontaneous closure in long-term follow up of small type VSD was 75%. They also reported the incidence of spontaneous closure in muscular type as 71.8% and in perimembranous 45%. Also these researchers have reported the incidence of abnormal pulmonary hypertension as (4%), aortic valve insufficiency (16%) along with large VSD. These findings are similar with our study (4, 9).

In another study, 1075 newborns in Russia were studied after birth by echocardiography and the incidence of spontaneous closure of VSD was 88.6% in the first year, in preterm newborns this criteria was 100% versus 78.8% in term newborns. Theses researchers proved that spontaneous closure of VSD in newborns occurred in a large scale (10, 15). In our research, the patients were studied after the period of being newborns.

In Roos and colleagues' study, it is indicated that the patients with VSD who underwent surgical operation, had good prognosis in long term follow up. Nearly 92% of their subject patients were in class 1 regarding NYHA class. In other study, Wu and colleagues indicated that those patients who were affected by diseases like aortic valve insufficiency, left ventricular stretch and pulmonary hypertension were in need of closed surgical operation for closure of VSD. Also in other study Gu and colleagues showed that it was possible to close the remaining VSD successfully after operation by intervention method and also these authors emphasized that there were no need for further surgery after closure of VSD. In the present research surgical operation was done in order to repair VSD for patients with large VSD, patients with aortic valve insufficiency after VSD, as well as patients with VSD and pulmonary hypertension. Although, these days VSD can be repaired by intervention method (6, 16 and 17).

In other research by Heuvel and colleagues, it is indicated that clinical course of patients with VSD was good in adulthood and in 6% of the patients the VSD had been closed spontaneously. Also no death has been occurred to any of the patients. 1.8% patient with endocarditis and 4% also due to hemodynamic problems underwent surgical operation. Also they indicated that VSDs with small defects have good pre-information in the future and are not in need of surgical operation and have no remarkable side effects. In our study, long term follow up of side effects was similar to other studies but bacterial endocarditis was less in this research (18).

In long term follow up, totally 4.34% of patients were affected by serious complications. In 28.2 % of the patients who underwent surgical operation the VSD remained in form of a small defect that did not require surgical intervention. Except for these cases: in 14 patients Eisenmenger syndrome was occurred due to delay in surgical operation time, In 2 cases, the further defect was repaired by surgical operation due to non-closure of VSD, and one case affected by stricture of pulmonary valve after surgical repair of VSD for whom, first echocardiography was performed, then angiography was performed in order to determine the severity of stricture and finally the cases were introduced to the surgeon for removal of the stricture. Among total, 3 patients with small VSD, were affected by aortic valve insufficiency that required VSD closure and valve repair. Also 3 cases were affected by endocarditis that was treated by antibacterial medical therapy and 2 patients with Eisenmenger underwent surgical operation with no complaints after 10 year follow up with normal pulmonary artery pressure in the recent catheterization. Also this study indicated

that in 54% of patients the defect was closed spontaneously before 36 months and in 10.2% of the cases surgery was closure of VSD, 43% of VSD cases the defect was closed over 36 months and in 17.9% the closure of VSD required surgical operation.

In recent years, the intervention method for closure of VSD (perimembranous and muscular types) has been employed in wide range as an alternative method. The study of Zheng and colleagues indicated that closure of VSD by percutaneous method under echocardiography had been useful and was associated with fewer complications. Because of shorted duration of hospitalization and less remaining scars, this method is more acceptable than open heart surgery. Also this method may decrease the incidence of heart block and aortic valve insufficiency with less morbidity (19 and 20).

Spontaneous closure of large VSD in present research was 0.6%, and in 98.7% this defect required surgical operation. The rest of the cases with large VSD could not undergo surgical operation due to the occurrence of the Eisenmenger syndrome. It is worth mentioning that other cases of VSD (53%) were under medical therapy and follow up, that this situation had occurred more than other centers, since a large number of patients refused to undergo surgical operation due to lack of financial capability .The long term complications in this study were similar to other studies (434%) except that small defects remained after surgical operation (28.32%)

Conclusions:

In this study, it is indicated that spontaneous closure of small VSDs will increase by time and a large number of moderate defects convert to small defects by time and treatment. Also a few numbers of large defects convert to small defects by time. The long term complications in this study were similar to other studies.

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