



## Original article

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Ljiljana Pejcić<sup>1</sup>, Bojko Bjelaković<sup>1</sup>,  
Biljana Miljković<sup>2</sup>,  
Radmila Mileusnić<sup>1</sup>, Gordana Grujić<sup>3</sup>,  
Ivica Pejcić<sup>4</sup>

<sup>1</sup>Clinic of Pediatrics,  
Clinical Center Nis

<sup>2</sup>Clinic of Gynecology,  
Clinical Center Nis

<sup>3</sup>Primary Health Care Center Nis

<sup>4</sup>Clinic of Oncology,  
Clinical Center Nis

## THE NATURAL HISTORY OF MUSCULAR VENTRICULAR SEPTAL DEFECTS

### SUMMARY

Ventricular septal defect (VSD) is the most frequently diagnosed congenital heart defect. The prognosis is usually good as it has spontaneous closure evolution, especially small muscular VSDs. The aim of this study was to determine the natural history of isolated muscular VSDs including frequency of spontaneous closure in relation to location in the muscular septum and age at the time of closure.

Doppler color flow mapping studies were performed to define ventricular septal defect anatomy, location and evolution. Of approximately 19,500 live births in the region during last 6 years, VSD was identified as isolated congenital heart lesion in 87 infants (incidence 5.8 per 1,000 live births). Forty-six infants (53%) had a muscular VSD. Thirty of 46 were followed up for a minimum of 1 year.

Spontaneous closure occurred in 17 of 30 cases (56.7%). The time of spontaneous closure ranged from 1 to 42 months and it was most commonly recorded during the first 6 months after birth. In the 6th month, 1st year and 18th month, spontaneous closure occurred in 11 (36.7%), 14 (46.7%) and 16 (53.3%) cases, respectively. It was registered in all cases except one within the first 18 months; the other defect closed in the 42nd month. It was remarkable that spontaneous closure was seen in 15 of 21 cases (71.4%) with apical VSD and 2 of 7 cases (28.6%) with trabecular VSD.

In conclusion, the frequency of spontaneous closure is very high in the first 6 months, especially within the first four years of life. Because of the high closure rate of muscular VSDs especially apical ones, it is recommendable to detect them early using color flow imaging and follow up patients up to spontaneous closure.

**Key words:** muscular ventricular septal defects, rate of spontaneous closure, color flow imaging

### INTRODUCTION

Ventricular septal defect (VSD) is the most common congenital cardiac anomaly encountered after bicuspid aortic valve. Isolated VSD occurs in approximately 2-6 of every 1000 live births (1.5-3.5 per 1000 term infants and 4.5-7 per 1000 premature infants) (1) and constitutes over 20% of all

congenital heart diseases. VSDs are slightly more common in females; 44% occur in males, and 56% occur in females. An area of residence may influence the prevalence of known VSDs. For example, a small muscular VSD is more likely to be defined in urban locations possibly because of greater access to sophisticated health care.

Since 1979, real-time 2-dimensional echo-

cardiography has dramatically improved the non-invasive anatomical assessment of VSD. Cross-sectional echocardiography coupled with Doppler echocardiography and color flow imaging can be used to determine the size and location of virtually all VSDs. (2, 3). In muscular septal defect, all views that image the ventricular septum must be employed. Color Doppler echocardiography is critical to determine small asymptomatic defects(4).

The evolution of the VSD has been the focus of several studies. The natural history has a wide spectrum, ranging from spontaneous closure to congestive cardiac failure and death in early infancy. Spontaneous closure of VSD especially in the first years of life is a well-known phenomenon and it occurs in about one third of all cases. Closure is most frequently observed in muscular defects (80%), particularly apical, followed by perimembranous defects (35-40%)(2).

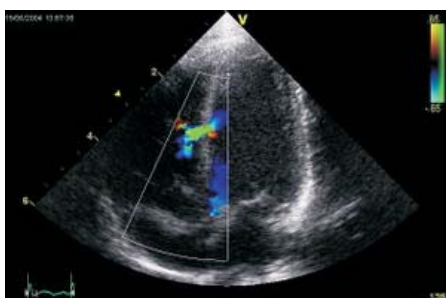
We followed up all patients with a muscular VSD, diagnosed over 6 years, to determine the frequency of spontaneous closure in relation to location in the muscular septum and age at time of closure.

#### MATERIAL AND METHODS

Of approximately 19.500 live births in the region under study (Nis and the surroundings) during last 6 years, VSD was identified as isolated congenital heart lesion in 87 infants using color flow Doppler echocardiographic examination (incidence 5.8 per 1,000 live births). Forty-six (53%) had a muscular VSD. Thirty of 46 were followed up for a minimum 1 year.

Echocardiographic examination was performed using available echocardiographic equipment (Hewlett Packard Image Point). Two dimensional, CW Doppler and color flow Doppler echocardiographic images were obtained at the standard parasternal long-axis view, classic and modified short-axis views and apical, subcostal four-chamber views. (Figure 1)

Figure 1. Color flow signals crossing the interventricular septum at the middle portion of the right ventricle show the presence of a small muscular ventricular septal defect on an apical four-chamber view.



When color imaging showed interventricular shunting, the diagnosis was confirmed by continuous and/or pulsed Doppler analysis, which indicated the timing and direction of the flow transversing the interventricular septum.

The muscular defects were categorized as apical, trabecular, or outlet, according to the classification of Gatzoulis et al (5). Defect sizes were measured in two-dimensional image or as the maximum thickness of color jet at the level of interventricular septum. VSDs were seemed large if the defects were as large as or greater than the aortic orifice, and small if only seen in some parts of the cardiac cycle or not seen at all but identified on color flow mapping. All other defects were classified as moderate.

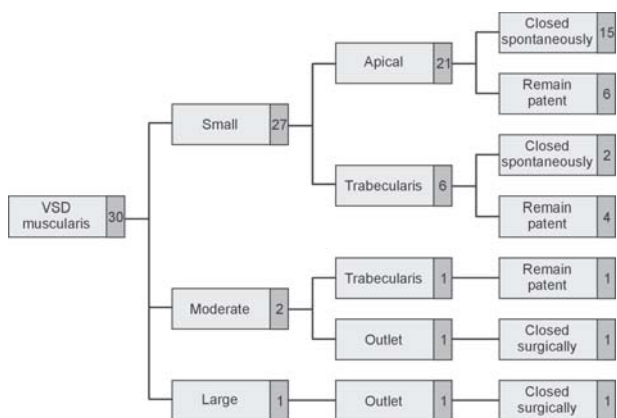
The patients were follow-up in intervals of 1, 3, and 6 months and 1 year. All received prophylaxis for infective endocarditis.

#### RESULTS

The ages of 30 cases who were followed-up for at least 6 years ranged from 1 day to 5 years old (mean  $5.23 \pm 7.03$  months) at the time of initial examination. Eighteen of 30 cases (60%) were diagnosed in the neonatal period. In 7 cases, initial echocardiographic examination was performed between the 1<sup>st</sup> and 6<sup>th</sup> month, in 3 cases between the 6<sup>th</sup> and 12<sup>th</sup> month, and only in 2 patients it was after the 12<sup>th</sup> month. Sixteen of them were female and 14 male (F/M=1.1). Patients were followed up to a minimum of 1 year of age and a maximum of 5 years.

Figure 2 summarizes the natural history of the VSDs.

Figure 2. The natural history of the 30 ventricular septal defects (VSDs) studied



Twenty seven VSDs were small, 21 of which were apical and 6 trabecular; two were moderate, one of which was outlet and one trabecular; and one was a large outlet VSD. Two outlet VSDs required surgical

closure including one moderate and one large. Of 28 patients managed non-surgically, 17 muscular defects spontaneously closed: 15 were apical and 2 were trabecular. Of 11 muscular VSDs that did not require surgical closure and remain open, 6 are apical and 5 are trabecular.

Spontaneous closure occurred in 17 of 30 cases (56.7%). The time of spontaneous closure ranged from 1 to 42 months and it was most commonly recorded during the first 6 months after birth. In the 6<sup>th</sup> month, 1<sup>st</sup> year and 18<sup>th</sup> month, spontaneous closure occurred in 11 (36.7%), 14 (46.7%) and 16 (53.3%) cases, respectively. This was seen in all cases except 1 within first 18 months; the other defect closed in the 42<sup>nd</sup> month. (Table 1)

Table 1. Time of Spontaneous Closure During Follow-up Period

Time of the spontaneous closure (months)	Number of patients	Ratio (%)	Cumulative ratio (%)
≤ 1	1	5.9	5.9
1 - ≤3	4	23.5	29.4
3 - ≤6	6	35.3	64.7
6 - ≤12	3	17.7	82.4
12 - ≤18	2	11.7	94.1
18 - ≤42	1	5.9	100.0
Total	17	100.0	-

It was remarkable that spontaneous closure was seen in 15 of 21 cases (71.4%) with apical VSD and 2 of 7 cases (28.6%) with trabecular VSD.

There was no record of infective endocarditis in any patient.

## DISCUSSION

A spontaneous closure is the most exciting aspect of the natural history of ventricular septal defect. All data in the literature point to its frequency. However, this frequency varies greatly from one study to another, depending on the population, age studied, follow-up period and the percentage of different types of VSD. In previous clinical studies, the rate of spontaneous closure of muscular VSD has been reported to be between 24% and 96%. These rates are quite different, but as a common result, most of the small defects close within few months after birth<sup>1(2,6)</sup>.

Some investigators suggested that small defects are not a malformation and that early spontaneous closure of these defects is a normal developmental process<sup>(7)</sup>.

Our results were partly predictable as we expected the increased spontaneous closure rate of muscular defect, especially apical. There are a few clinical reports related to the rate of closure for muscular VSDs and influence of location on spontaneous closure VSD. Ramaciotti et al. (8) reported that the rate of closure for muscular VSDs and apical muscular VSDs was 24% and 23%, respectively. They emphasized that spontaneous closure of muscular VSDs was most commonly seen in the first 18 months of life. They also observed that the natural history of single muscular VSD is not influenced by location in the muscular septum.

Du et al. (7) screened full-term neonates with color flow Doppler imaging for muscular VSDs. The rate of closure at the end of the first year was 84.8%, but only one-fourth of defects were located in the apical region. They found that defects localized in the apical region and defects >4 mm in size remain patent more than VSDs located elsewhere.

Hiraishi et al (4) found a very high frequency for isolated VSDs when term neonates were routinely investigated using echocardiography. Most of the defects were small and muscular and 76% had closed by the age of 1, but 45% were apical muscular VSDs.

Turner et al. (6) confirmed that the position of a ventricular septal defect is extremely relevant to its natural history. The spontaneous closure rate for muscular defects was significantly greater than in perimembranous defects. Shirali (2) and colleagues studied 156 cases whose mean age was 28 months and also found a significantly higher spontaneous closure rate for muscular defects.

Our findings are very similar to those reported by Turner et al.<sup>2</sup> and Atalay et al. (9). In Atalay's study, a very high frequency of spontaneous closure of apical muscular VSDs was found. Spontaneous closure was seen in 24 of 42 cases (57.1%) between 1 and 36 months of age, and it was most commonly recorded during the first 6 months of life. Sapin et al. (10) found very high closure rate (75%) for apical VSDs, too.

Spontaneous closure becomes less likely during adolescence and adult life. In the study by Gabriel et al (11), spontaneous closure occurred in 6% of patients.

## CONCLUSION

A high chance of spontaneous closure is one of the major reasons why small VSDs are followed conservatively. However, diagnosing even a small VSD is important because of the risk of infective endocarditis. It is necessary to follow up patients to determine the spontaneous closure, especially within

the first years of life. Reported closure rates vary with size and location of VSD, age at presentation and patient population. Small VSDs have a >50% chance of spontaneous closure by four years of age.

Because of the high closure rate of muscular VSDs, especially apical, and the absence of serious clinical signs, parental anxiety should be minimized.

## REFERENCES

1. Mehta AV, Chidambaram B. Ventricular septal defect in the first year of life. *Am J Cardiol* 1992; 70: 364-366.
2. Shiralli GS, O'Brian Smith E, Geva T. Quantitation of echocardiographic predictors of outcome in infants with isolated ventricular septal defect. *Am Heart J* 1995; 130: 1228-1235.
3. Hornberger LK, Sahn DJ, Krabill KA, et al. Elucidation of the natural history of ventricular septal defects by serial doppler color flow mapping studies. *J Am Coll Cardiol* 1989;13: 1111-1118.
4. Hiraishi S, Agata Y, Nowatari M, et al. Incidence and natural course of trabecular ventricular septal defect; two dimensional echocardiography and color doppler flow imaging study. *J Pediatr* 1992; 120: 409-415.
5. Gatzoulis MA, Li J, Ho SY. The echocardiographic anatomy of ventricular septal defects. *Cardiol Young* 1997; 7: 471-484.
6. Turner SW, Hunter S, Wyllie JP. The natural history of ventricular septal defects. *Arch Dis Child* 1999; 81: 413-416.
7. Du ZD, Roguin N, Wu XJ, et al. Spontaneous closure of muscular ventricular septal defect identified by echocardiography in neonates. *Cardiol Young* 1998; 8: 500-505.
8. Ramaciotti C, Vetter JM, Renee A, et al. Prevalence, relation to spontaneous closure and association of muscular ventricular septal defects with other cardiac defects. *Am J Cardiol* 1995; 75: 61-65.
9. Atalay S, Tutar E, Ekici F, Nacar N: Spontaneous closure of small apical muscular ventricular septal defects. *The Turkish Journal of Pediatrics* 2005; 47: 247-250.
10. Sapin SO, Junkel PA, Wong AL, et al. The congenital apical ventricular septal defect. *Pediatrics* 1994; 93: 516-518.
11. Gabriel HM, Heger M, Innerhofer P, et al. Long-term outcome of patients with ventricular septal defect considered not to require surgical closure during childhood. *J Am Coll Cardiol*, 2002; 39: 1066-1071.

## PRIRODNA ISTORIJA MUSKULARNIH VENTRIKULARNIH SEPTALNIH DEFEKATA

Ljiljana Pejčić<sup>1</sup>, Bojko Bjelaković<sup>1</sup>, Biljana Miljković<sup>2</sup>, Radmila Mileusnić<sup>1</sup>, Gordana Grujić<sup>3</sup>, Ivica Pejčić<sup>4</sup>

<sup>1</sup> Klinika za pedijatriju Kliničkog centra u Nišu

<sup>2</sup> Klinika za ginekologiju i akušerstvo Kliničkog centra u Nišu

<sup>3</sup> Dom zdravlja Niš

<sup>4</sup> Klinika za onkologiju Kliničkog centra u Nišu

### SAŽETAK

Ventrikularni septalni defekt (VSD) najčešća je urođena srčana mana. Prognoza je u najvećem broju slučajeva dobra, posebno ako se radi o malim muskularnim defektima, imajući u vidu njihovu sklonost ka spontanom zatvaranju. Cilj ovoga rada bio je da utvrdi prirodnu evoluciju izolovanih muskularnih VSD-a, odnosno frekvencu spontanog zatvaranja u zavisnosti od njihove lokacije u muskularnom septumu, kao i od uzrasta bolesnika.

Anatomija, lokacija i evolucija defekata praćena je korišćenjem Color Doppler tehnike. Za proteklih 6 godina, od približno 19500 živorođene dece u Nišu i okolini, VSD kao izolovana srčana mana nađena je u 87-oro (incidenca 5.8%). Njih 46-oro (53%) imalo je muskularnu lokalizaciju defekta. Tridesetoro dece, od njih 46, kontinuirano je praćeno najmanje po godinu dana.

U 17 od 30-oro dece došlo je do spontanog zatvaranja VSD-a (56.7%). Vreme spontanog zatvaranja kretalo se u intervalu od 1 meseca do 42 meseca po rođenju, pri čemu se najveći broj njih desio u prvih 6 meseci. Na kraju 6. meseca bilo ih je 11 (36.7%), na kraju 1. godine 14 (46.7%), a na kraju 18. meseca ukupno 16-oro dece (53.3%) sa spontano zatvorenim VSD-om. Dakle, samo kod jednog deteta zatvaranje defekta registrovano je posle 18. meseca, tj. u 42. mesecu života. Treba istaći da je čak u 15 od 17 (71.4%) defekt bio lociran u apikalnom delu muskularnog septuma, dok je samo 2 od 17 (28.6%) imalo trabekularni VSD.

Može se zaključiti da se najveći broj spontanih zatvaranja odvija u prvih 6 meseci života, odnosno do 4. godine. Pri tome se najčešće zatvaraju apikalni muskularni VSD-i, te ih treba na vreme otkrivati i kontinuirano pratiti do njihovog zatvaranja.

**Ključne reči:** muskularni ventrikularni septalni defekti, spontano zatvaranje, Color Doppler