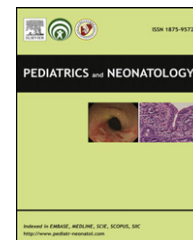


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ORIGINAL ARTICLE

Color Doppler Echocardiographic Study on the Incidence and Natural History of Early-Infancy Muscular Ventricular Septal Defect

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Key Words

color Doppler echocardiography; muscular ventricular septal defect; natural history

Background: Most small muscular ventricular septal defect (M-VSD) types have been diagnosed using color Doppler echocardiography. The purpose of this study was to understand the incidence of small M-VSD in the neonatal period and analyze the natural history of these M-VSDs. **Materials and Methods:** All individuals in our study were neonates delivered at term who had a normal healthy appearance. Each accepted neonate had an examination with complete color Doppler echocardiography once before discharge. If the examination was confirmed for M-VSD, the study participants were then classified according to defect type. Further examination was arranged with color Doppler echocardiography at 1 month, 2 months, 4 months, 6 months, 9 months, and 12 months of age or until there was complete spontaneous closure.

Results: Among 2891 neonates, we found that 72 (24.9/1000) were diagnosed with M-VSD. Among this group, 38 were male and 34 were female. Only six infants were lost to follow-up. Fifty-four of the 66 infants (81.8%) had M-VSD closed spontaneously at 12 months' follow-up. Significantly, 33 of 37 infants (89.2%) with mid-muscular type, the most common type of M-VSD, closed within the 1st year of life compared with apical type (17/24:70.8%). Four of the five infants (80%) had anterior type M-VSD closed. Infants with posterior type M-VSD were not seen during this study period.

Conclusion: Although the incidence of M-VSD was common in the neonatal period, there was also a high rate of spontaneous closure. Therefore, comparison of M-VSD appearance with the incidence of congenital heart disease in neonates had a decisive influence on analysis. Copyright © 2011, Taiwan Pediatric Association. Published by Elsevier Taiwan LLC. All rights reserved.

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1. Introduction

Most muscular ventricular septal defect (M-VSD) presentations are small and clinically asymptomatic; cardiac murmurs cannot easily be heard in these neonates. Most defects do not need any treatment and have a high rate of spontaneous closure. If not for careful examination in the neonatal period, most M-VSDs would not be diagnosed, and the incidence rate would also be underestimated.^{1–12} Recently, color Doppler echocardiography has been widely used in newborn screening, resulting in early diagnosis of clinically insignificant small M-VSDs and also indirectly enhancing the incidence of neonatal period congenital heart disease (CHD). However, there has been scant literature found discussing the rate of spontaneous closure in different subtype of M-VSD within the 1st year of life.¹³ Therefore, we designed this study, hoping to understand the incidence of small M-VSD in neonatal period and analyze the impact of natural history of the defects.

2. Methods

This study focused on full-term live normal newborn births, defined as 37–42 weeks of pregnancy and birth weight 2500–4200 g, with no obvious abnormalities, chromosomal disorders, or infectious diseases at nursery between January 1994 and December 1995. Parental consent was obtained for each neonate. Color Doppler echocardiography was routinely performed to clearly check the complete ventricular septum. If a newborn was diagnosed with an M-VSD, examinations were arranged with complete color Doppler echocardiography at 1 month, 2 months, 4 months, 6 months, 9 months, and 12 months respectively, coordinating together with the childhood vaccination schedule. The diagnosis of M-VSD was established on the basis of two criteria, including the presence of a mosaic image passing through the muscular ventricular septum from the left to the right ventricle (Figure 1) and a turbulent systolic flow jet recorded on the right surface of the VSD by pulsed or continuous wave Doppler (Figure 2). Care was taken to avoid misdiagnosing the accelerated flow in the right ventricular trabeculae or coronary artery as a VSD.¹⁴ All examinations were recorded on videotape and

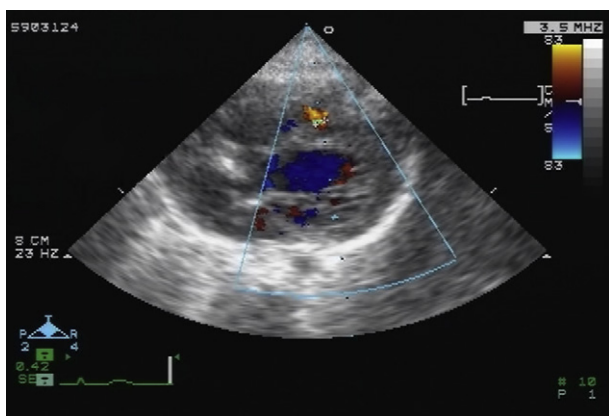


Figure 1 Mosaic image of blood flow passing through the muscular ventricular septum from the left to the right ventricle.

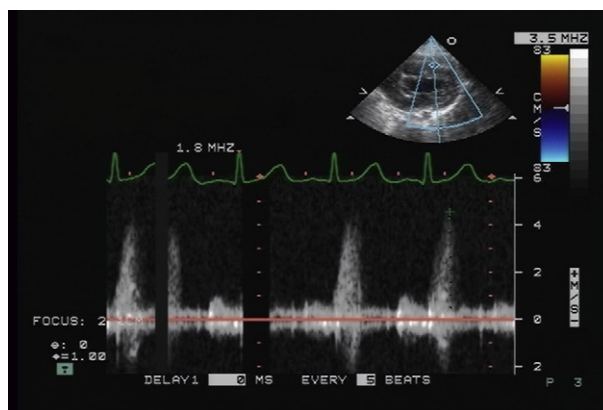


Figure 2 Turbulent systolic flow jet recorded on the right surface of the ventricular septal defect by continuous wave Doppler flow tracing.

reviewed by at least two pediatric cardiologists. According to Kirklin et al,¹⁵ echocardiography findings in infants with an M-VSD can be classified as anterior, mid-muscular, apical, and posterior, and changes in its natural course can be statistically analyzed. The anterior muscular defects are located anterior to the septal band, which extends along the mid-septum from the insertion of the moderator band toward the membranous septum. Mid-muscular defects are posterior to the septal band, anterior to the septal attachment of the tricuspid valve, and superior to the moderator band. Posterior muscular defects are located posterior to the septal attachment of the tricuspid valve. Apical muscular defects are located inferior to the moderator band. The Friedman test was used to test the difference of closure rate for different types of M-VSD. The statistical significance level was p value less than 0.05.

3. Results

Among the 2891 newborns examined by complete Doppler echocardiography, a total of 72 cases were diagnosed as

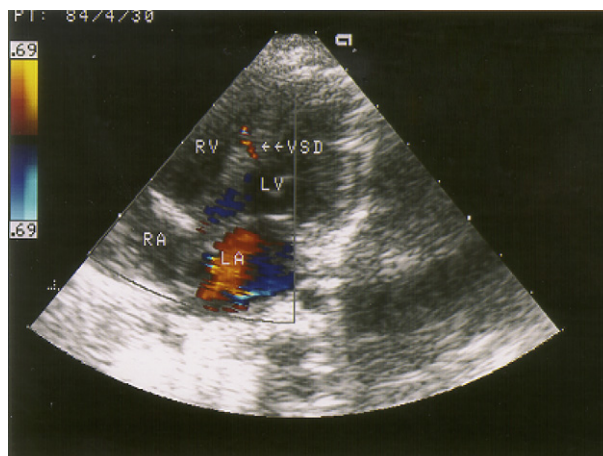


Figure 3 Mosaic image of blood flow passing through the muscular ventricular septum from the left to the right ventricle, indicating mid-septal type M-VSD. M-VSD = muscular ventricular septal defect.

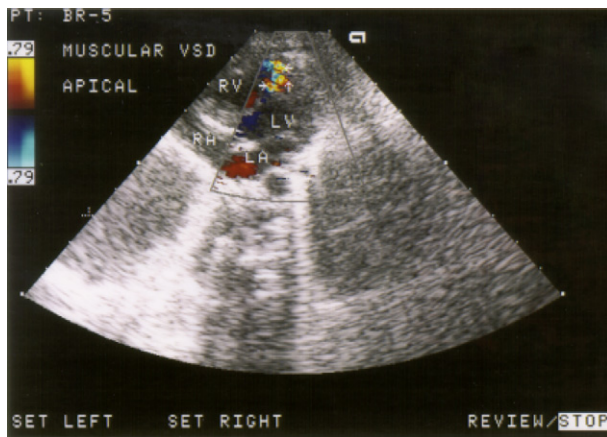


Figure 4 Multiple turbulent systolic flow jets recorded on the apical region of the ventricular septal defect, indicating apical type M-VSD. M-VSD = muscular ventricular septal defect.

M-VSD; the incidence was 24.9 per 1000 live births (72/2891). From these 72 cases, 66 cases were accepted, as 6 infants were lost to follow-up. At 1-year follow up, complete closure was noted in 54 cases (81.8%). According to the defect location, 33 of 37 cases (89.2%) were of the mid-muscular type (Figure 3) and 17 of 24 cases (70.8%) were of the apical type (Figure 4) that had closed completely. However, the anterior M-VSD type had the lowest incidence rate (only 5 cases), but four of the cases closed completely (80.0%). Infants with posterior M-VSD type were not seen during this study period (Table 1). Because most of M-VSDs were small in size, we could not evaluate the correlation between the size of the M-VSD and the timing of spontaneous closure. During more than 1 year of tracking, four cases of Swiss cheese-type apical M-VSD did not close spontaneously. All of the six cases with multiple defects had not spontaneously closed yet, including the four cases of Swiss-cheese type and two cases with multiple defects in the mid-septal region. It was also found that 12 cases of M-VSD had not closed spontaneously during follow-up until the present. The Friedman test was used to test the difference of closure rate in three types of M-VSD. The *p* value for Friedman test was 0.012 (Figure 5).

4. Discussion

The most common form of CHD in childhood is VSD, occurring in 50% of all children with CHD; of the VSDs, 20% are isolated lesions.^{16,17} According to past statistical

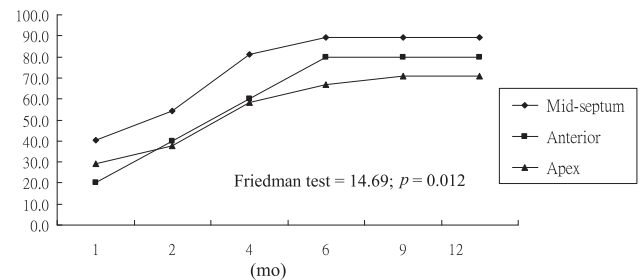


Figure 5 A graph documenting the percentage of time and closure rate for different types of muscular ventricular septal defect.

analysis, the incidence of isolated VSDs in neonates was 1.5–2.5/1000.^{18–20} However, a recent report by Kapusta et al²¹ found that the incidence was gradually increasing. Hiraishi et al¹³ reported that live-birth neonates had a risk of about 4.46 per 1000 for VSDs; however, they also reported VSD rates as high as 2% of live-birth neonates. Some authors have recently reported higher incidences of VSD when they carried out neonatal screening by echocardiography.^{13,14,22,23} They concluded that the incidence of VSD may have been previously underestimated. Increased preterm delivery, environmental factors, or better echocardiographic imaging because of evolving technology may be responsible for the increase in the reported incidence of VSD.²⁴ Most likely, many of these muscular VSDs are smaller defects diagnosed in newborn infants that might have been previously undetected before the routine use of color Doppler echocardiography. Furthermore, studies have suggested that the prevalence of VSD would be even greater if echocardiography were universally performed rather than only on infants with suspected CHD.^{14,25,26}

The VSDs can be classified into four types: membranous, muscular, endocardial cushion, and subpulmonary. The previous data found that membranous defects were the most common type of VSD^{27,28}; however, a recent study reported that by using color Doppler echocardiography in neonatal examination, the incidence of muscular ventricular defects was not lower than those of membranous types.⁸ This was comparable with other reports and still higher than the incidence of other types of VSDs. Many factors could have contributed to this significant difference. Some authors agreed that most of these M-VSDs in the neonatal period were difficult to diagnose, especially apical septum defects or other very small defects, which can now be examined by color Doppler echocardiography.^{13–15,18–21,27–34}

Spontaneous closure is the most exciting aspect of the natural history of M-VSDs. Although these rates are quite

Table 1 Closure rate of each subtype of muscular ventricular septal defect.

Type	Follow-up time						
	Newborn	1 mo	2 mo	4 mo	6 mo	9 mo	12 mo
Mid-septum	37 (0)	15 (40.5)	20 (54.1)	30 (81.1)	33 (89.2)	33 (89.2)	33 (89.2)
Apex	24 (0)	7 (29.2)	9 (37.5)	14 (58.3)	16 (66.7)	17 (70.8)	17 (70.8)
Anterior	5 (0)	1 (20.0)	2 (40.0)	3 (60.0)	4 (80.0)	4 (80.0)	4 (80.0)

Data are presented as *n* (%).

different, most of the small defects close within few months after birth.⁶ Hiraishi et al¹³ found a very high frequency of isolated VSDs when term neonates were routinely investigated using echocardiography. Most of the defects were small and muscular (76% had closed by the age of 1), but 45% were apical M-VSDs. Du et al³⁵ screened full-term neonates with color flow Doppler imaging for M-VSDs and found that the rate of closure at the end of the 1st year was 84.8%, but only one-fourth of the defects were located in the apical region. Thus, in neonates, it was difficult to know the exact incidence and natural history of the defects if there were no immediate examination with color Doppler echocardiography. Color Doppler echocardiographic examination was performed by clinicians to confirm the diagnosis of neonatal-period CHD only when infants were found to have heart murmur, respiratory distress, tachycardia, or a failure to thrive. Most of the small M-VSDs closed spontaneously at 6 months of age; thus, if the neonates were asymptomatic or without heart murmur, their small M-VSDs would not be examined by color Doppler echocardiography, underestimating their true incidence. Major factors influencing the spontaneous closure rate of VSDs include the patient's age at the first examination, localization and size of the defects, and length of the follow-up period.¹⁰

In our study, spontaneous closure was seen in 81.8% cases of M-VSDs during the 1st year of life, occurring most commonly in the mid septum. Despite the increased incidence of VSD, most patients' VSDs were clinically insignificant, and the spontaneous closure rate was remarkably high within the 1st year of life. Color flow Doppler echocardiographic screening is the most reliable method for assessing the true incidence of VSD and can detect VSD in neonates, even in cases without murmurs.²⁴

5. Conclusion

Although there was a high spontaneous closure rate in M-VSDs, long-term regular follow-up might be necessary to detect accurate timing of the closure in different parts of defects. According to the data from our own and previous studies, we also can further trace the approach and treatment guidelines for infants with M-VSDs.

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