Therapeutic strategy for multiple VSD combined with MVSD in infants and young children

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Introduction

As a kind of special ventricular septal defects (VSD), the multiple VSD is usually exhibited as the perimembranous VSD combined with the muscular ventricular septal defect (MVSD), the VSD in other sites (subpulmonary VSD or atrioventricular canal VSD), malignant VSD, combined with MVSD or simply multiple MVSD. Surgical intervention is usually applied to the multiple VSD, MVSD presenting a challenge for the limitations in various kinds of surgery methods, e.g. surgical suture, sandwich suture, suture via a small incision in left ventricular, hybrid closure or simple intervention closure. This is a retrospective cohort study analyzing the treatment method for the MVSD of the multiple VSD, which could serve as a reference for the selection of more rational treatment methods.

Patients and Methods

Patients

This study was conducted on 63 pediatric patients admitted to our hospital with multiple VSD between January 2010 and April 2013 to perform surgery or transcatheter closure. The patients were 33 males and 30 females. The research team obtained the approval of the Ethics Committee of our hospital and informed consents of the children patients’ guardians. The age of patients was between 6-28 months (10 ± 6) months and weighed between 5.5 and 18.0 (7.1 ± 2.9) kg. Primary repair was performed for all of the patients; the MVSD in 7 patients was not detected during the surgery and no extra treatment was taken. The surgical suture was performed for 36 patients, hybrid repair under the direct vision for 8 patients, and hybrid repair via the right ventricle for 7 patients. After surgery, we followed up the patients regularly to reexamine the X-ray image of the chest, EEG, and color Doppler echocardiography to observe the closure of MVSD and the presence of a residual shunt.

RESULTS: All of the 63 enrolled patients with multiple VSD survived without perioperative death. Three patients who were undergoing hybrid repair under direct vision received delayed sternal closure. One patient who was undergoing hybrid repair under direct vision had a postoperative cardiac dysfunction. 55 patients were followed up for 1 to 24 months. 28 patients had residual shunt of varying degrees during the follow-up, and most of the MVSD of patients with residual shunt was less than 4 mm, who were receiving further follow-up and observation.

CONCLUSIONS: Appropriate surgical strategies can be applied according to the specific surgical condition for the treatment of MVSD in infants with multiple VSD, and the satisfactory clinical outcome can be achieved.

Key Words: Multiple VSD, MVSD, Hybrid closure, Residual shunt.
3rd and 4th ribs in the left chest, with chest x-ray showing an enlarged heart, ECG showing ventricu-
lar hypertrophy on both side and Color Dop-
pler echocardiography suggesting multiple VSD. Preoperative diagnosis was made, with 38 cases of perimembranous VSD combined with MVSD, 6 of subpulmonary VSD combined with MVSD, 1 of atioventricular canal VSD combined with MVSD and 5 of simply multiple VSD. Out of the total of 63 cases, 31 cases combined with atrial septal defect (ASD) and 14 cases combined with patent ductus arteriosus (PDA) were both combined with no other complex cardiac anomalies.

Treatment Methods
Surgical treatment was done under combined in-
travenous and inhalation anesthesia with endotrache-
al intubation. The chest was opened in the middle of the sternum and the cardiopulmonary bypass (CPB) was established. After blocking the canal vein, the cold blood cardioplegia was perfused at the aortic root. Once the heartbeat stopped, MVSD was detected through an incision of the right atrium. Then, dacron pad intermittent suture was applied to 14 cases of MVSD. MVSD was clipped through the left and right ventricles using sandwich method with two Gore-Tex meshes with a diameter larger than 2 to 3 mm in 18 cases; 4 cases of MVSDs were repaired through the restricted small incision of left ventricle with Gore-Tex, MVSDs in 8 cases were blocked through the placement of umbrella-shaped tap under the di-
rect vision in the operation, and no treatment was conducted for 7 cases whose MVSD could not be de-
tected. Hybrid repair via the right ventricle with heart beating was performed in 7 cases under the guidance of transesophageal echocardiography (TEE) before the establishment of CPB; transcatheter closure was directly performed under the guidance of TEE for the simply multiple VSD in 5 cases.

Follow-up
The patients were followed from 1 to 23 months after surgery. Reexamination by X-ray image of the chest, EEG, and color Doppler echocardiography was performed immediately after surgery and 1, 3 and 6 months after surgery, to observe the closure of MVSD and the presence of a residual shunt.

Table I. Statistics of basic clinical data of enrolled patients.

<table>
<thead>
<tr>
<th>Cases</th>
<th>Age (months)</th>
<th>Weight (kg)</th>
<th>HR (beats/min)</th>
<th>SPO₂ (%)</th>
<th>Surgery duration (min)</th>
</tr>
</thead>
<tbody>
<tr>
<td>63</td>
<td>10.0±6.0</td>
<td>7.1±2.9</td>
<td>112.2±28.7</td>
<td>98.3±1.2</td>
<td>186.4±59.4</td>
</tr>
</tbody>
</table>

Statistical Analysis
Statistical software (Version X; IBM, Armonk, NY, USA) was utilized for statistical analysis. Student’s t-test was adopted for the intergroup comparison of two samples and X²-test for the constituent ratio of two samples. Measurement data were presented as mean ± standard deviation (x ± SD) and the count data as a percentage (%). p<0.05 was considered statistically significant.

Results
Basic Clinical Data of Enrolled Patients (x ± s)
In this study, there were 33 males and 30 fe-
males. The patients aged from 6 to 28 months with an average of (10 ± 6) months and were weighed 5.5 to 18.0 kg with an average of (7.1 ± 2.9) kg. Basic clinical data of patients were listed in Table I.

Postoperative Follow-up of the Echocardiogram Indexes
We followed up the postoperative condition of patients from 1 to 23 months with an average of (18.4 ± 8.7) months. No death occurred in the pe-
rioperative period. After surgery, 6 cases of patients delayed the closure of chest with MVSD closed in the direct vision and 3 cases of patients showed increased left ventricular end diastolic diameter (LVEDD) and decreased left ventricular ejection fraction (LVEF) compared with the preoperative LVEDD and LVEF, which suggested statistically significant differences (p<0.05). In addition, no significant cardiac dysfunction was found in other patients after surgery, and compared to the preo-
perative LVEDD, the postoperative LVEDD was significantly less than the preoperative with statisti-
cally significant differences (p<0.05). See Table II.

Comparison of the Levels of Cardiac Markers of Enrolled Patients Before and After Surgery
After surgery, through the follow-up on the car-
diac markers in the serum of the enrolled patients, we found that the levels of creatine kinase (CK-MB), cardiac troponin (cTnI) and pro-brain natri-
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uretic peptide (pro-BNP) after the surgery were significantly lower than those before the surgery with statistically significant differences ($p<0.05$). See Table III.

**The Follow-up of the Enrolled Patients**

In the follow-up of 63 patients, we failed to detect the MVSD in 25 cases, and identified 7 patients with MVSD (1 case with the diameter of MVSD larger than 6 mm being followed up to perform the second time of surgery, 1 case with the diameter of MVSD larger than 4 mm, and 5 cases with the diameter of MVSD less than 4 mm). In 14 patients who MVSDs were sutured intermittently using Dacron pad, there were 6 with residual shunt and 8 with no presence of a residual shunt. In 18 patients whose MVSDs were clipped using the sandwich method, the residual shunt was not seen in 15 patients but 3 patients were found to be with filamentous residual shunt (diameter < 1 mm). In 4 patients whose MVSDs were repaired through the restricted small incision of the left ventricle with Gore-Tex, only 1 patient was with filamentous residual shunt (diameter < 1 mm). In 8 cases whose MVSDs were blocked through the placement of umbrella-shaped tap under the direct vision in the operation, residual shunt (diameter < 4 mm) in varying degrees was found in 3 patients and no residual shunt was found in the other 5 patients. In 7 patients who accepted hybrid repair via the right ventricle for MVSDs, we found that 2 patients were with the residual shunt (diameter < 4 mm). In the 5 patients who accepted the direct transcatheter closure of multiple VSD, 2 patients were found with filamentous residual shunt (diameter < 4 mm). All the patients were still under the follow-up and no mortality was observed.

**Discussion**

Due to the large shunt volume of VSD from the left ventricle to the right ventricle, pediatric patients with multiple VSD are prone to clinical symptoms manifested by shortness of breath, hyperhidrosis, susceptibility to the respiratory tract infection, even the recurrent pneumonia, cardiac dysfunction, and retarded or stunted growth and development, which requires intervention at an early stage. The MVSD, usually seen in pediatric patients with multiple VSD, is difficult to handle due to the large anatomic variances. Currently, various treatment methods are available for MVSD, which include surgeries, hybrid repair and intervention closure. However, the surgical methods for MVSD, including the incisions through the left and right ventricles and ventricular muscle incision, can lead to the postoperative cardiac dysfunction, various arrhythmia and apical aneurysm for patients. In this study,
we performed surgical treatment for the MVSD of 36 patients, in which the MVSDs of 14 patients were sutured intermittently using two pads, and the MVSDs of 18 patients were repaired using the sandwich method, and the MVSDs of 4 patients were sutured through the incision of the left ventricle with Gore-Tex meshes. The results showed that cardiac dysfunction was identified in 3 patients after intermittent suture surgery, with residual shunt diameter being < 3 mm in 6 cases and 4.6 mm in 1 case. For the patients in whom the sandwich method was performed, filamentous residual shunt was found in 3 patients and the rate of residual shunt was significantly lower than those in the former methods, indicating that sandwich method is the more reliable surgical treatment for correcting the MVSD and the worth to be applied in the clinical practice. In 4 patients who accepted the suture through the incision of the left ventricle with the Gore-Tex mesh, no significant left ventricular dysfunctions were observed, despite there was 1 case with residual shunt. In recent years, direct vision hybrid repair through the right ventricle and transcatheter closure have become the new treatment options for MVSD. In this study, out of 8 pediatric patients who accepted hybrid closure in the direct vision residual shunt with a diameter less than 4 mm was identified in 3 patients and no shunt was seen in 5 patients, suggesting that the application of hybrid closure is reliable and effective for the MVSD. There was delayed closure of chest cavity and cardiac dysfunction in 2 patients, indicating that cardiac dysfunction might be more susceptible to the treatment for the MVSD using hybrid closure in the direct vision, especially placing multiple VSD occlusion devices; the possible reason was that the occlusion devices can affect the movement of ventricular wall to a certain degree. Hybrid closure through the right ventricle was performed for 7 patients, in which we did not find any migration of the occlusion device in the follow-up or cardiac dysfunction after surgery, and residual shunt was presented in 2 patients, one of which had a residual shunt with a diameter of 4.5 mm, also suggesting that the hybrid closure through the right ventricle is safe and reliable with gratifying efficacy. Transcatheter closure is a widely accepted procedure among physicians and internist as there is no need to establish a CPB and has a good prognosis. According to the literature, it was reported that occluder device was displaced in 1 patient with apical MVSD. In this study, we performed the transcatheter closure under the guidance of TEE for 5 patients with simply MVSD, and found that residual shunt with a diameter less than 3 mm was found in 2 cases and no displacement of occluder device was observed, suggesting the safety and effectiveness of this method. Besides, we found that 6 patients with MVSD all survived without cardiac dysfunction as no treatment was given to them. Among these patients, 4 cases showed residual shunt, in which 3 cases with a diameter less than 2 mm and 1 case with a diameter of 4.9 mm. It indicated that the treatment is not necessary for MVSD with a diameter of about 4 mm, which is coincident with the results of the literature. Besides, we also noticed that MVSD was not found in the follow-up after surgery in 2 patients, indicating a larger possibility of misdiagnosis of MVSD by color Doppler echocardiography. Despite the various therapeutic strategies for MVSD, each method has its strengths and shortages. For the simply multiple MVSD, transcatheter closure should be considered; for the independent MVSD with a larger defect, sandwich method for the closure of MVSD will be considered, and the intermittent suture by using two pads is recommended for the MVSD with a smaller defect. For those with a high occurrence rate of residual shunt, the application of the suture with the pad through a restricted incision of the left ventricle is suggested. Though the hybrid closure in a direct vision has been proved to be relatively efficient, multiple occlusion devices have required the enclosure of multiple MVSD, leading to an increase in the medical cost and the possibility of delayed closure of chest, as well as the cardiac dysfunction of child patients after surgery. Hybrid closure through the right ventricle can avoid the establishment of CPB and significantly reduce the occurrence of cardiac dysfunction, but there remains the possibility of ectopic; thus, it is suitable for the apical MVSD, which is difficult to be observed. Due to the self-healing ability, no treatment is required for the MVSD with a diameter of about 4 mm and less significant influence on the hemodynamics and which can hardly be successfully treated by hybrid closure through the right ventricle.

Conclusions

Individualized therapy should be applied to the muscular defect of the infant multiple VSD. For the simple multiple MVSD without any other anomalies, we can consider the transcatheter closure,
for the MVSD explored in the operation, we can apply the surgery or hybrid closure in a direct vision according to the sites and size. Also, for the MVSD unable to be explored in the operation, we can attempt to perform the hybrid closure through the right ventricle under the guidance of TEE after surgery, but if the attempt fails, no treatment is required for the MVSD with a diameter of about 4 mm. Thus, rational selection of therapeutic strategy can achieve better efficacy in the treatment of multiple VSD.

Conflict of interest
The authors declare no conflicts of interest.

References