

## Is Left Ventriculotomy Feasible for Muscular Ventricular Septal Defects in Infants?

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### ABSTRACT

Optimal management of muscular ventricular septal defects (MVSDs) remains controversial. Left ventriculotomy is the cornerstone of surgical repair but is frequently complicated by residual shunts, left ventricular dysfunction, apical aneurysm, or arrhythmias. In this study, we evaluated the long-term outcomes of surgical repairs in infants with isolated MVSDs. We retrospectively analyzed clinical data from 56 children with MVSDs (31 males, 25 females). Follow-up by questionnaire and Doppler echocardiography was performed at discharge and between 2 and 124 months after surgery. Patient age was 2 to 40 months (median, 21 months) and weight was 3.0 to 15.3 kg (median, 5.3 kg). Two patients died after surgery (hospital mortality, 3.57%). One patient with MVSDs died of low cardiac output caused by the long duration of cardiopulmonary bypass. Another patient with Swiss cheese MVSD received a single patch closure but died of low cardiac output immediately after cardiopulmonary bypass. Immediate complications such as a third-degree atrial-ventricular block occurred in 2 patients, but they recovered before discharge and showed no residual shunt. No deaths occurred during follow-up, but a residual shunt was found in 1 patient. Delayed complete heart block requiring a pacemaker occurred in 1 patient. One patient showed paroxysmal supraventricular tachycardia that was treated with amiodarone. The left ventricular ejection fraction was 0.45-0.55 in 8 patients and 0.55-0.73 in 46 patients. No apical aneurysm was found. All the surviving patients returned to normal school life. Our results indicate that surgery is a suitable treatment option in infants and children with isolated MVSDs and that preoperative diagnosis is crucial to a successful outcome. Infants can

tolerate a left ventriculotomy incision for MVSDs in the lower or apical ventricular septum.

### INTRODUCTION

Optimal surgical management of muscular ventricular septal defects (MVSDs) in infants remains controversial despite options such as initial pulmonary artery banding, single-stage repair with a transatrial or a transventricular approach, interventional transcatheter device closure [Becker 2004], or a hybrid strategy with an intraoperative patch or periventricular device closure techniques [Diab 2005]. Although surgery is common for most patients with MVSDs, each technique is associated with significant mortality and complications.

Novel surgical approaches, such as through a tricuspid valve, a perimembranous VSD, or the interatrial septum, may improve outcomes. However, MVSDs in the lower or apical intraventricular septum require left ventriculotomy, which has a long-term association with left ventricular dysfunction, apical aneurysm, or arrhythmias when performed in pediatric patients [Wollenek1996]. We retrospectively reviewed the clinical data to determine immediate and long-term surgical outcomes in 56 infants with MVSDs in the apical septum who received surgical repairs via left apical incisions.

### PATIENTS AND METHODS

#### Patients

We retrospectively reviewed the clinical data for 56 patients who underwent surgical treatment for MVSDs during the period from April 1997 to March 2007 at the Department of Cardiovascular Surgery, Xijing Hospital, the Fourth Military Medical University (Xi'an, China). The study was approved by the Institutional Ethics Committee of Xijing hospital. The parents or guardians of all patients provided informed consents. There were 31 boys and 25 girls. Patient age ranged from 2 to 40 months (median, 21 months) and weight from 3.0 to 15.3 kg (median, 5.3 kg). In 48 patients, pansystolic harsh murmurs were appreciated in the left sternal border along with a fine systolic thrill; in 8 patients, heart murmurs presented only after administration of vasoactive

October 29, 2007; Received in revised form January 31, 2008; accepted February 8, 2008.

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drugs (nitroprusside sodium 0.3-0.5  $\mu\text{g/kg}$  per minute or prostaglandin E 5-20  $\text{ng/kg}$  per minute) for 1 week or more. Nine patients showed cyanosis after exercise. All patients had pulmonary congestion and a dorsoventral cardiothoracic ratio of between 0.60 and 0.64. Electrocardiogram results were normal in 33 patients and demonstrated right ventricular hypertrophy in 23 patients. In 15 of these patients the right ventricular hypertrophy was associated with a right bundle-branch block. Preoperative ultrasonic echocardiogram showed mild or moderate pulmonary hypertension (30-75 mm Hg) in 32 patients, but all shunts were primarily left-to-right before surgery.

An MVSD was defined as an intraventricular communication with a rim consisting entirely of muscle. MVSDs were characterized by preoperative echocardiographic imaging and intraoperative inspection. A Swiss cheese septum was defined as a highly deficient ventricular septum with 4 or more MVSDs. Characteristics of the MVSDs in the study patients are shown in the Table.

## OPERATIVE TECHNIQUES

Cardiopulmonary bypass (CPB) was established with moderate hypothermia (28°C-32°C) by standard aortic and bicaval venous cannulation. The left ventricle was decompressed by a vent placed through the interatrial septum. Myocardial protection was antegrade cold crystal cardioplegia.

Perimembranous VSD required a right atriotomy for transatrial repair and viewing the tricuspid valve. Subpulmonary and infracristal VSDs were closed through the right atrium and the tricuspid valve, or through the pulmonary artery. MVSDs smaller than 5 mm were closed by a direct interrupted suture with 4-0 or 5-0 polypropylene monofilament (Premilene, B/Braun, Germany). MVSDs larger than 5 mm were repaired by use of autologous pericardium that had been fixed in 0.4% glutaraldehyde and closure with a running suture.

MVSDs were identified by gentle probing with right-angled pliers through the tricuspid valve to delineate a connection with the left ventricle, even through a perimembranous VSD or the interatrial septum. Because all patients in this study had at least 1 MVSD in the apical or lower intraventricular septum, a left apical incision (carp-shaped mouth) parallel to the anterior descending artery was used for repairs and

viewing. Because of few muscular bundles, right-angled pliers could be used to identify the connection from the left to right ventricle. In some patients, however, more than 1 opening in the right ventricle could connect with the opening in the left ventricle, requiring careful probing. Defects were closed with patches or direct sutures except in 1 patient with Swiss cheese defects that required a large Dacron patch.

After VSDs were closed, the vent was removed temporarily and the left apical incision was closed in a sandwich method with 2 polyester felt patches. The lung was inflated and air was evacuated from left atrium, then the atrial septum was closed. Before the ascending aorta was unclamped, the lung was inflated to look for residual shunts. Ejection of florid oxygenation blood from the right ventricle indicated the presence of other left-to-right shunts.

Postoperative echocardiograms performed in all patients before discharge were retrospectively reviewed by a single, blinded cardiologist. Follow-up data were obtained from clinic records when available or from written correspondence with patients' parents, information that is continuously updated in our surgical database. Median follow-up time was 6.4 years (range 3 months to 10.2 years).

## RESULTS

Two patients died after surgery (hospital mortality, 3.57%). In 1 patient, after weaning from CPB, systolic murmurs were audible by auscultation, and in this patient we repaired 2 MVSDs (2 mm and 3 mm) directly with pledgeted sutures after resuming CPB. However, the patient died of low cardiac output due to lengthy CPB. Another patient had a large subarterial VSD (10 mm) and 4 MVSDs (7 mm, 8 mm, 5 mm, and 5 mm). Although the MVSDs were closed with a large Dacron patch on the left ventricular side, the patient died of low cardiac output after CPB removal. Third-degree atrioventricular blocks occurred in 2 patients, both of whom recovered before discharge. The median CPB duration was 131 minutes (range, 72 to 190 minutes) and the median cross-clamp time was 71 minutes (range, 39 to 103 minutes).

No deaths occurred during the follow-up period of between 4 months and 124 months. One patient had a residual shunt and moderate pulmonary hypertension (65 mm Hg), with a VSD in the muscular septum. One patient who had presented with a third degree atrial-ventricular block and recovered at discharge showed a complete heart block 16 months after surgery, requiring a permanent pace maker. One patient showed paroxysmal supraventricular tachycardia that was treatable with amiodarone, and 15 patients showed right bundle-branch block. The left ventricular ejection fraction was 0.45-0.55 in 8 patients and 0.55-0.73 in 46 patients. No apical aneurysms were found. All surviving patients returned to normal activities including attending school.

## DISCUSSION

Ideal treatment in infants with MVSDs would incorporate a single-stage repair, complete VSD closure, and avoidance of arrhythmias and ventricular dysfunction. A left ventriculotomy

Table. Characteristics of Multiple Ventricular Septal Defects (MVSDs) in the Study Patients (N = 56)

A. Location	No. of MVSDs
Posterior	31
Anterior	37
Midtrabecular	46
Apical	68
B. No. of MVSDs per Patient	No. of Patients
2	15
3	18
4	17
5	6

provides intraoperative exposure and allows for complete repair, especially with MVSDs located in the lower intraventricular or apical septum. The use of this technique is limited, however, because it may also impair long-term cardiac function and cause arrhythmias.

We used a left ventriculotomy incision to perform surgical correction in 56 patients with MVSDs in the apical intraventricular septum. Operation mortality was 3.57%, with no subsequent deaths. Transient atrioventricular block occurred in 2 patients, and 1 patient underwent recurrent complete heart block. Two patients died of low cardiac output despite the administration of vasoactive agents. No apical aneurysms were seen, and 1 patient had a residual shunt and 1 had paroxysmal supraventricular tachycardia. Surviving patients resumed normal activities, including school attendance. Overall, our results indicate that left ventriculotomy can be used successfully, with acceptable morbidity and mortality, in infants undergoing isolated MVSD repair procedures.

Historically, identification of MVSDs required cardiac catheterization and left ventriculography, with an 86% discovery rate. Currently, MVSDs can generally be diagnosed by Doppler echocardiography alone [Spevak 1993], except in cases with pulmonary artery stenosis, pulmonary hypertension, short-haul defects, and/or superior position infundibular or nonrestrictive VSD. We treated 2 patients with undiagnosed MVSDs; 1 died due to prolonged CPB and the other had pulmonary hypertension and a residual shunt. The results in these cases indicate that preoperative or intraoperative detection of MVSDs shortens operative time and decreases the residual shunt incidence.

Patients with MVSDs develop pulmonary hypertension and right heart failure and should be treated rapidly after diagnosis. Right ventricular and muscular septum hypertrophies increase the incidence of residual shunts. Therefore, preventing residual shunts is key for a successful outcome. Adequate MVSD exposure can be improved by selecting the correct cardiac incision, with right atrial or ventricular incisions chosen in superior or central VSDs, and low incisions in cases of right ventricular dominance. Low intra-VSDs might require other approaches because of coarse trabeculations of the right ventricle, and a left ventriculotomy may be necessary. In another study, 9 patients, including 1 neonate who underwent surgery during the hebdomadal period, received VSD repair through left ventricular incisions and had normal left ventricular ejection fractions at 47 months of follow-up [Kitagawa1998]. Serraf [1992] reported, however, that left ventriculotomy for apical lesions, but not left ventricular dominance, increased the risk of both recurrent VSD and death because the incision led to left ventricular dysfunction. In our study, however, we found normal cardiac output in all but 2 patients after left ventriculotomy, despite poor cardiovascular function in the same patients preoperatively.

Cardiac conduction disturbances and ventricular arrhythmias are potential surgical complications in the patients we describe. Before surgery, 15 of our patients (26.8%) had a right bundle-branch block due to right ventricle hypertrophy. After surgery, a right bundle-branch block and a left anterior fascicular block were associated with moderator band

resection or the placement of a surgical MVSD patch. Left ventriculotomy could also lead to scar formation and produce arrhythmias. We found no arrhythmias in our study patients, however, in part because the surgical field view provided by the ventriculotomy prevented further ventricular muscular resection. We also found no residual shunts during long-term follow-up, presumably because we inflated the lung to check for residual shunts before the ascending aorta was opened.

Novel approaches to treat MVSD are being developed. For example, Alsoufi et al [2006] reported that reendocardialization enables early complete or nearly complete obliteration of MVSDs with minimal residual lesions (shunt, ventricular dysfunction). Between 1986 and 1991, Lecal et al [1994] used fibrin glue to repair MVSDs in 15 children, and color-flow mapping in the 13 long-time survivors revealed only minimal residual shunts. The procedure they used, however, sometimes causes systemic embolization of fibrin glue particles. In addition, Lecal et al did not evaluate prospective efficacy, and the study was limited by low numbers of patients. Macé et al [1999] used a large single patch in the repair of Swiss cheese VSDs, a technique that eliminates the need to identify individual MVSDs at the defect site. We also used a single-patch closure in the patient with the Swiss cheese lesion, but the patient died of low cardiac output immediately after surgery. In neonates and small infants, this technique may cause postoperative cardiac dysfunction because the large patch restricts the motion of the ventricular septum wall. For the other 4 patients in our group who had Swiss cheese defects, we identified all MVSDs by left ventriculotomy and repaired them with direct pledgeted sutures, and all patients recovered. Joshua et al [2006] reported that Amin et al used periventricular closure with an Amplatzer device for VSD occlusion, and the technique was updated by Bacha et al with excellent results in the first series of patients studied, but patients experienced long-term complications such as residual shunts, cardiac conduction abnormalities (right bundle branch block, left anterior fascicular block), and ventricular arrhythmias.

## CONCLUSION

MVSDs are sometimes straightforward congenital heart anomalies that are simple to treat, but complex treatment may be required for multiple defects or defects situated in the lower or apical ventricular septum. Other techniques for treating these patients have been attempted, and surgical management is still the treatment of choice for multiple septal defects. For MVSDs in the lower or apical ventricular septum, postoperative ventricular performance can be improved by vasoactive agents, and a left ventriculotomy can be tolerated when allowing for residual shunts.

## REFERENCES

- Alsoufi B, Karamlou T, Osaki M, et al. 2006. Surgical repair of multiple muscular ventricular septal defects: the role of re-endocardialization strategy. *J Thorac Cardiovasc Surg* 132(5):1072-80.
- Becker P, Frangini P, Heusser F, et al. 2004. New surgical approach to device closure of multiple apical ventricular septal defects. *Ev Esp Cardiol* 57(12):1238-40.

- Diab KA, Hijazi ZM, Cao QL, et al. 2005. A truly hybrid approach to perventricular closure of multiple muscular ventricular septal defects. *J Thorac Cardiovasc Surg* 130(3):892-3.
- Joshua D.C. Robinson, Frank J, et al. 2006. Cardiac conduction disturbances seen after transcatheter device closure of muscular ventricular septal defects with the Amplatzer occluder. *Am J Cardiol* 97(4):558-60.
- Kitagawa T, Lucian LA, Mosca RS, et al. 1998. Techniques and results in the management of multiple ventricular septal defects. *J Thorac Cardiovasc Surg* 115:848-56.
- Leca F, Karam J, Vouhe PR, et al. 1994. Surgical treatment of multiple ventricular septal defects using a biologic glue. *J Thorac Cardiovasc Surg* 107:96-107.
- Macé L, Dervanian P, Le Bret E, et al. 1999. "Swiss cheese" septal defects: surgical closure using a single patch with intermediate fixings. *Ann Thorac Surg* 67(6):1754-9.
- Serraf A, Lacour Gayet F, Bruniaux J, et al. 1992. Surgical management of isolated multiple ventricular septal defects: logical approach in 130 cases. *J Thorac Cardiovasc Surg* 103:437-42.
- Spevak PJ, Mandell VS, Colan SD, et al. 1993. Reliability of Doppler color flow mapping in the identification and localization of multiple ventricular septal defects. *Echocardiography* 10:573-81.
- Wollenek G, Wyse R, Sullivan I, et al. 1996. Closure of muscular ventricular septal defects through a left ventriculotomy. *Eur J Cardiothorac Surg* 10:595-8.