Surgical Closure of Apical Muscular Ventricular Septal Defects via a Right Venticulotomy

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Identification and surgical closure of apical muscular ventricular septal defects are associated with certain morbidity and mortality because of their location in the ventricular septum distal to the moderator band, making adequate visualization and complete closure from the right atrium almost impossible. However, the precise margins of apical VSDs can be readily identified by a small apical right ventriculotomy. We report two infant patients with congenital heart disease and apical muscular VSDs communicating between the left ventricular apex and the RV infundibular apex. Patient A was a 9-month old female who had a swiss-cheese type VSD (apical VSDs below the moderator band and one VSD at the high infundibular portion). Patient B was a 18-day old male who had coaractation of the aorta (CoA), patent ductus arteriosus (PDA), and multiple VSDs (subarterial doubly committed type VSD with apical muscular type VSDs). Their apical VSDs were closed with separate patches via apical right ventriculotomy. The associated cardiac defects were all corrected during the single operation. Good biventricular contractility without residual VSD were confirmed by 14-month follow-up. The successful outcome of these two patients indicates that surgical closure of apical muscular VSDs can be achieved safely and completely in early infancy through a limited apical right ventriculotomy.

Key Words: Apical VSD • Right ventriculotomy • Apical right ventriculotomy

INTRODUCTION

Apical muscular ventricular septal defects (VSDs) occur between the left ventricular apex and the infundibular apex, rather than between the left and right ventricular apices. These defects are often complex, compromising several openings on either side of the septum and an oblique and tortuous intraseptal course.1,2 Small muscular defects without hemodynamic significance often close spontaneously.3 Large muscular defects can lead to pulmonary hypertension, pulmonary edema, respiratory insufficiency, and failure to thrive. The most common methods for managing apical VSDs are right atriotomy, left ventriculotomy or palliative therapy by pulmonary artery banding; however, all are associated with significant limitations. For example, closure of the apical muscular ventricular septal defect via right atriotomy through the tricuspid valve is often not possible when the defect is located far from the tricuspid valve plane, distal to the moderator band, because it is hidden by multiple muscular trabeculations which prevent the surgeon from identifying the anatomical borders of the defect. Closure via left ventriculotomy can solve that problem; however an incision on the left ventricular myocardium can lead to apical left ventricular aneurysms or dyskinesis later in life. The disadvantages of pulmonary artery banding are numerous, including early operative mortality. Therefore, a safer and more effective approach is necessary.2,4-6 The precise margins of apical muscular VSDs can be readily identified via a small apical right ventriculotomy (Figure 1).
We present two infant patients with congenital heart disease combined with apical muscular VSDs communicating between the left ventricular apex and the RV infundibular apex. The apical VSDs in both patients were repaired via an apical right ventriculotomy (Figure 1). Patient A had a swiss-cheese type VSD (apical VSDs below the moderator band and one VSD at the high infundibular portion) (Figure 2 & Figure 3); patient B had coarctation of the aorta, patent ductus arteriosus (PDA), and multiple VSDs (subarterial doubly committed-type VSD with apical muscular-type VSDs).

Patient A was 9 months old and weighed 8 kg (5th percentile); patient B was 18 days old, and weighed 3 kg (5th percentile). Patient A presented with mild to moderate degree of pulmonary artery hypertension, Pp/ Ps: 0.43, Qp/Qs > 4, Rp/Rs: 0.1, and cardiomegaly with C/T ratio: 0.62. Patient A had had progressive tachypnea since birth, and presented with cardiomegaly with C/T ratio: 0.64. Electrocardiograms of the two patients before the operations both revealed biventricular hypertrophy. Patient A underwent a median sternotomy and the apical muscular VSD was initially repaired via right atriotomy; however the apical muscular VSD was not clearly visible after the tricuspid valve was retracted, requiring the surgeon to make three unsuccessful attempts to close the apical VSD. Intraoperative transesophageal echocardiogram (TEE) revealed that significant VSD flow between the LV and the RV persisted without any change in the size of the defect. A longitudinal incision (15-20 mm) was made through the right ventricle apical free wall, parallel to and to the right of the distal part of the left anterior descending coronary artery (Figure 1).

Through this small incision, the infundibular apical septal surface was clearly visible. Patient B underwent a median sternotomy. His subarterial-doubly-committed VSD was repaired via an RA-PA approach; the pre-
ductal coarctation of the aorta was repaired by coarctectomy, end-to-end anastomosis, and PDA ligation. The apical muscular VSD was repaired via an apical right ventriculotomy.

RESULTS

The associated cardiac defects in both patients were all corrected during the respective single operations. The two patients were weaned from cardiopulmonary bypass easily. The cardiopulmonary bypass time of patient A was 171 minutes; patient B was 237 minutes. Aortic cross-clamp time for patient A was 60 minutes; patient B was 80 minutes. Post-operative transesophageal echocardiography (TEE) in both patients revealed small residual VSDs (1.5 mm in diameter) at the apical muscular area with restrictive blood flow. The subsequent postoperative hospital course was uneventful. Two-dimensional and color Doppler echocardiography before discharge confirmed good biventricular contractility with trivial residual VSD in both patients. The two patients were followed by our pediatric cardiologist; the trivial residual apical VSDs had closed spontaneously and both of them were clinically well at 14-month follow-up. Body weight percentile of the two patients increased from 5th percentile to 25th and 75th percentiles, respectively. The C/T ratio had decreased to 0.52 in both cases after operation. Postoperative EKG revealed no ventricular hypertrophy.

DISCUSSION

Morphologically, the right ventricle is composed of the right ventricular sinus, which is derived from the proximal part of the bulbus cordis, and the right ventricular infundibulum, which is derived from the distal part of the bulbus cordis. The junction between the right ventricular sinus and the infundibulum is formed by a ring of conal musculature consisting of the conal septum that extends out onto the parietal or free wall as part of the parietal band, and by the septal band and the moderator band. This infundibular ring demarcates and slightly separates the right ventricular sinus posteroinferiorly from the infundibulum anterosuperiorly. Apical muscular VSDs, which are commonly regarded as defects between the left and right ventricular apices, are in fact defects in the apical portion of the ventricular septum that separates the left ventricular apex from the infundibular apex. The infundibular apex is located anterosuperior and to the left of the apex of the RV sinus and constitutes the lowermost part of the infundibular chamber. This is observed in both D-loop and L-loop ventricles.

Successful closure of apical muscular VSDs is known to be difficult and frequently suboptimal. The approach to muscular ventricular septal defect has been controversial. The area of the ventricular septum separating the left ventricular apex from the infundibular apex — where as a rule, apical VSDs occur — is frequently not visible through the orifice of the tricuspid valve. The moderator band and the multiple trabeculations beneath it create a multiperforated wall that hides the apical muscular VSD from a transtricuspid view. Successfully repairing apical muscular VSDs is frequently impossible. Although apical VSDs are clearly visible via left ventriculotomy, the procedure can cause the formation ventricular aneurysm, and left ventricular apical dyskinesis. Furthermore, scar tissue at the incision level is often responsible for triggering arrhythmic episodes. Palliative pulmonary artery banding (PAB) is sometimes an unavoidable means to reduce the shunt temporarily; however, disadvantages of such banding are numerous, including stenosis, distortion of the peripheral pulmonary artery, persistent congestive heart failure or cyanosis. In each of our two patients with apical muscular VSDs, we performed a longitudinal incision of 15-20 mm into the right ventricle apical free wall, on the right of and parallel to the distal part of the left anterior descending coronary artery. This incision exposed a region of the right ventricular septal surface that is called the apex of the infundibulum and in which apical muscular ventricular septal defects are usually located. Through this incision, identification of the defects in both infants was extremely safe and easy. Although small residual apical VSDs were found in both patients after surgery, both residual apical VSDs had closed spontaneously by the first- and fourth-month follow-ups, respectively. No other complications were noted at the fourteenth-month follow-up. Most studies on RV function have regarded the RV as a single anatomic and functional unit and attempted to evaluate its function as one chamber. The RV, however, is composed
of several anatomic segments that can be divided into two major components: the RV sinus and the RV infundibulum. Data from embryological, anatomical, and molecular observations suggest that the RV sinus and the infundibulum are distinct chambers evolved from different parts of the embryonic heart. Little is known about regional differences between the RV sinus and the RV infundibulum with regard to their relative contribution to global RV systolic function. We know from the study by Geva, Powell, Crawford et al. that the RV sinus performs most of the pump function of the right heart and the infundibulum serves mostly as a pulsatile conduit, ejecting only a portion of the combined RV stroke volume. From this study, we concluded that apical right ventriculotomy is a safer method than left ventriculotomy.

CONCLUSION

The successful outcomes of these two patients indicate that surgical closure of apical muscular VSDs can be achieved safely and completely in early infancy through a limited right ventricular apical ventriculotomy. Long-term follow-up of these and similar patients is needed to provide further evaluation of this approach.

REFERENCES

經由右心室以手術方式關閉頂端部肌肉型心室中隔缺損

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以藥物或者是手術的方式治療多肌肉型心室中隔缺損在臨床上仍是有高的致病率以及死亡率。在手術過程中，經由右心房要去確認，進而以手術方式關閉位於遠在右心室肌肉群之後的肌肉型心室中隔缺損，在臨床上有很大的困難。不過，經由在右心室尖頂端部的一個小切口，去直接探視頂端部肌肉型的心室中隔缺損，在我們的經驗中，是非常容易的，而且可以很明確的看清楚缺損的範圍。

在 2004 年 5 月，有 2 位先天性心臟病病人併有頂端部肌肉型心室中隔缺損，此缺損位於左心室肌肉心室中隔的頂端以及右心室的漏斗部之間。病人甲是一位 9 個月大女生，其心臟缺損為乳酪型心室中隔缺損併有頂端部肌肉型心室中隔缺損，而病人乙則是一位 18 天大的男嬰，其缺損為多重肌肉型心室中隔缺損 (含動脈下心室中隔缺損以及頂端部的肌肉型心室中隔缺損) 及主動脈弓縮窄。

我們藉由經右心室尖頂端部切開的方式進行頂端部的肌肉型心室中隔缺損的修補。此二位病人，由同樣的方式完成手術，而完全的矯正了心室的結構異常。在術後心臟超音波追蹤下，僅剩餘微不足道的心室中隔缺損血流，而且仍具有良好的左右心室收縮功能。在術後第 5 個月的心臟超音波追蹤發現，之前剩餘心室中隔缺損已完全消失，而且沒有任何併發症。藉由這二個成功的例子，可以提供我們一些經驗，即使在嬰兒期，藉由右心室尖頂端部的一個小視野，進行頂端部肌肉型的心室中隔缺損補修是一個安全, 成功率高而且比較少後遺症的手術方式。

關鍵詞：肌肉型心室中隔缺損、右心室切開術。