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A Surgical Strategy for Cor Triatriatum Atrial Septal Defect—A1 Lam Subclass

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ABSTRACT

Atrial fibrillation and a heart murmur were diagnosed in a 68-year-old woman during a routine medical examination. She presented 2 years later with pulmonary edema. A transthoracic echocardiography examination revealed a tunneled atrial septal defect (ASD) and severe tricuspid regurgitation. Transesophageal echocardiography and 3-dimensional computed tomography evaluations revealed multiple intracardiac defects, including abnormal atrial septation suggestive of a typical cor triatriatum sinistrum (A1 Lam subclass), a rare congenital defect in adults. The patient underwent tricuspid valve repair with concomitant closure of the ASD by using the cor triatriatum curtain to form an autologous transposition flap. The intraoperative transesophageal and predischarge imaging evaluations confirmed an excellent repair. The patient made a swift recovery and demonstrated improvement in her symptoms at follow-up. This previously undescribed technique eliminates the need for a prosthetic implant and is applicable in >80% of cor triatriatum cases in which an ASD exists.

INTRODUCTION

Cor triatriatum sinistrum (CTS) is a very rare congenital cardiac malformation, especially in the older population. It is characterized by the presence of a fibromuscular membrane dividing the left atrium into 2 chambers. There are a number of different subtypes according to the Lam classification [Rodefeld 1990; Alphonso 2005]. The classic presentation in 80% of cases is associated with an atrial septal defect (ASD), but it may also be associated with other congenital malformations (atypical presentation), such as Ebstein anomaly, partial anomalous pulmonary venous connections, double-outlet right atrium, and double-outlet right ventricle [Rodefeld 1990]. The complexity of this anomaly may explain why CTS can frequently be misdiagnosed [Kaminishi 1999]. This report illustrates a previously undescribed surgical approach for closing a typical CTS of A1 Lam subclass.

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CASE REPORT

A 68-year-old woman was referred to the cardiac surgery outpatient clinic with progressive exertional dyspnea and angina. She had been hospitalized in an acute condition with pulmonary edema in the months preceding her surgical referral. Her exercise tolerance was limited by her cardiac symptoms to less than one-half mile on the flat at a reduced pace. A clinical examination revealed signs of significant tricuspid regurgitation, and the patient was in atrial fibrillation. Precordial examination revealed a systolic murmur. A transthoracic echocardiography (TTE) examination during her recent acute admission revealed the presence of a tunneled ASD (approximately 2.5 cm) associated with a left-to-right shunt and severe tricuspid regurgitation with gross annular dilatation (>7 cm). A coronary angiography evaluation revealed an unusually large circumflex coronary artery and a small, malpositioned left anterior descending artery. A transesophageal echocardiography (TEE) examination revealed concurrent intracardiac defects. Three-dimensional (3D)-reconstruction images from a cardiac computed tomography (CT) examination were suggestive of a cor triatriatum with an associated ASD and normal pulmonary venous connections (Figure 1).

The surgical approach was through the right atrium. A median sternotomy immediately revealed a grossly enlarged right atrium. The normal pulmonary venous connection was verified on inspection. After establishing cardiopulmonary bypass through bicaval venous cannulation and an ascending aortic return, we lowered the core temperature to 32°C. Myocardial protection was provided by antegrade cardioplegia. The right atrium was isolated between caval snares and was entered through a longitudinal incision. The tricuspid annulus was severely dilated. The cor triatriatum defect was identified through the ASD. The septal element of the cor triatriatum membrane was in direct continuity with the lower border of the ASD. The superior and inferior edges of the cor triatriatum septum were mobilized through the ASD to form an autologous flap large enough to close the defect (Figure 2). A structurally normal mitral valve became visible across the atrial defect through a widely open aperture. The autologous curtain already attached to the base of the ASD was sized, trimmed, and then sewn to the free edges of the atrial septum. A standard tricuspid annuloplasty ring was selected

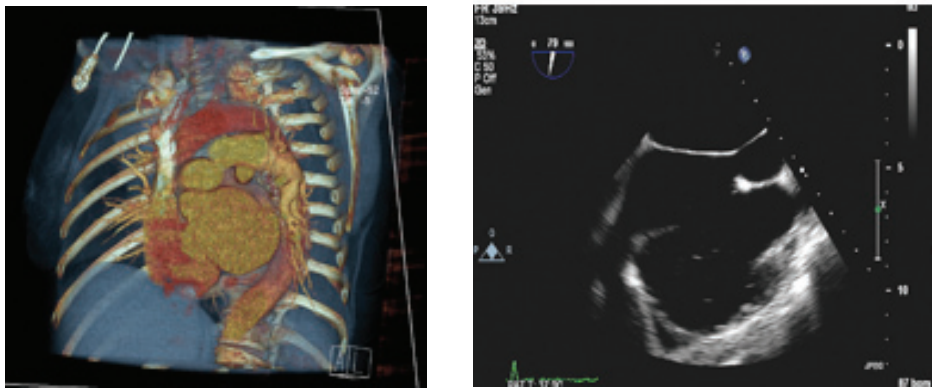

Figure 1. Three-dimensional reconstruction image (A) from a cardiac computed tomography examination (B) suggestive of cor triatriatum with an associated atrial septal defect and normal pulmonary venous connections.

after careful sizing and then secured to the native tricuspid annulus. The right atrium was then closed, and the left atrial appendage was excised to minimize future thromboembolic risk in the setting of chronic atrial fibrillation. The patient was weaned from cardiopulmonary bypass without difficulty. The intraoperative TEE examination confirmed both a satisfactory tricuspid repair with no residual regurgitation and elimination of the interatrial shunt. Sequential measurements of oxygen saturation in key cardiac chambers confirmed the absence of any significant step-up. The patient's postoperative recovery was uneventful. A routine predischarge TTE examination confirmed no residual ASD. At the 6-week follow-up, the patient remained asymptomatic with a greatly improved exercise tolerance—2 to 3 miles daily walking without stopping and no difficulty with uphill climbs.

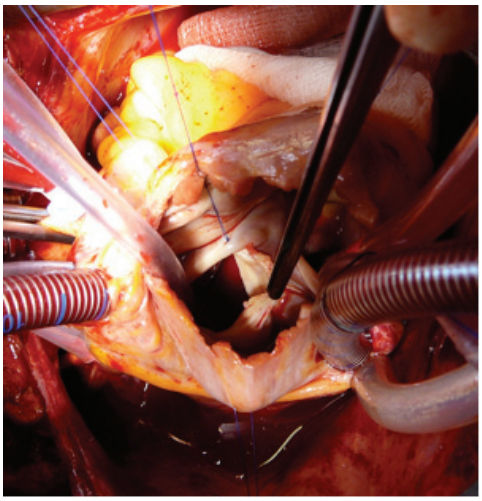

Figure 2. The superior and inferior edges of the cor triatriatum septum were mobilized through the atrial septal defect to form an autologous flap large enough to close the defect.

DISCUSSION

This report describes a typical CTS of the A1 Lam subclass [Alphonso 2005]. In this subclass, the proximal left atrial chamber receives all the pulmonary veins; the distal chamber contains the left atrial appendage and the mitral valve. There is an ASD between the right atrium and the proximal chamber. The diagnosis in this case was made preoperatively via TEE. Precise anatomic definition, however, was made possible with 3D imaging techniques.

The objective of this report is to describe the successful use of an autologous left atrial curtain for surgical repair of an ASD in the setting of CTS. The number of alternative closure techniques that have been described include use of bovine pericardium [Kaminishi 1999], autologous pericardium [Lâcis 2004; Ozkökeli 2007], GORE-TEX patch [McLean 2008], and Dacron felt [Das 2006]; however, transposition of an autologous left atrial membrane to close an ASD has not heretofore been reported for this congenital anomaly. This technique eliminates the need for a prosthetic implant and is applicable in >80% of cases of cor triatriatum in which an ASD coexists.

The histologic makeup of the atrial membrane has been studied extensively [Van Praagh 1969]. It consists of the wall of the common pulmonary vein dorsally and the wall of the left atrium ventrally. Therefore, the composition of the membrane adjacent to the left atrial cavity is that of the atrial free wall. External to this membrane are layers of striated left atrial myocardium and a layer of fibrous tissue corresponding in part to the left atrial epicardium. The bilaminar muscular structure of the cor triatriatum membrane therefore is evidently a strong autologous material. Its continuity to the interatrial septum would preserve blood supply to the flap. This material may have advantages, especially in pediatric practice with respect to septal growth as the patient's heart grows. A free graft may be equal with respect to retaining its tensile strength conferred by its fibrous constituent.

CONCLUSION

CTS is an extremely rare congenital anomaly in adults. It may present with symptoms of progressive cardiac failure

in the absence of coronary disease. The diagnosis of this condition requires multimodality investigations, including advanced 3D imaging. In the majority of cases, it is associated with an ASD. In these cases, the cor triatriatum membrane may be used as a transposition flap for closing the intra-atrial defect. This approach has not previously been descried. The use of an autologous material may prove advantageous in pediatric settings. The immediate assessment of this approach proved that it was a success. The patient's short-term outcome was remarkable in terms of symptom relief. Long-term follow-up of this patient is anticipated.

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