



Case report

Surgical repair of perimembranous ventricular septal defect and aortic regurgitation in an adult patient with Laubry-Pezzi syndrome.

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Abstract

Laubry-Pezzi syndrome is a congenital heart disease that consist in a prolapse of aortic valve cusping into a subjacent ventricular septal defect due to Venturi effect. It results in progressive aortic valve insufficiency. The perimembranous type is the most common due to the proximity of the aortic annulus to such defects. The aim of this report is to highlight the specificity of the diagnosis and the surgical management of this syndrome in adult patients.

Keywords:

Laubry-Pezzi syndrome; aortic regurgitation; ventricular septal defect; surgery.

Introduction

Laubry and Pezzi first described the association of ventricular septal defect (VSD) and aortic regurgitation (AR) in 1921. The syndrome is a congenital heart insidious disease. The management of this rare pathology is still non-consensual regarding operative timing and techniques. The early diagnosis of the VSD is capital before the appearance of AR. Few cases of Laubry-Pezzi syndrome have been reported in the literature mostly in children [1,2]. we report a case of Laubry-Pezzi syndrome in an adult patient managed in our cardiovascular surgery department.

Observation

We report the case of 32-year-old man patient with a previously asymptomatic ventricular septal defect (VSD) presented with New York Heart Association class II symptoms and signs of severe aortic insufficiency. The echocardiography showed perimembranous ventricular septal defect VSD 7mm and severe aortic regurgitation. The right ventricular (RV) WAS 52 mmHg denoting of pulmonary hypertension (figure 1). The Laubry Pezzi syndrome was diagnosed and surgery was indicated.

A median sternotomy incision was performed. The Cardiopulmonary bypass was established using bicaval cannulation, and moderate hypothermia. After aortic cross clamping, the ascending aorta was opened through an oblique incision allowing an access in an "trans-aortic approach". The intraoperative findings were confirmed by inspecting the aortic sinuses and elevating the right coronary cusp. There was a perimembranous VSD and prolapsed aortic valve in the right coronary cusp. We closed the VSD using interrupted, pledgeted horizontal mattress sutures (figure2). The procedure is completed by a free margin placating of the right coronary cusp. This reduction of the free edge of the right coronary cusp elongated and repositioned the hinge point of the right coronary cusp to restore the normal height of the cusp, hence a normal surface of apposition. The procedure was verified by a post bypass intraoperative transesophageal echocardiography.

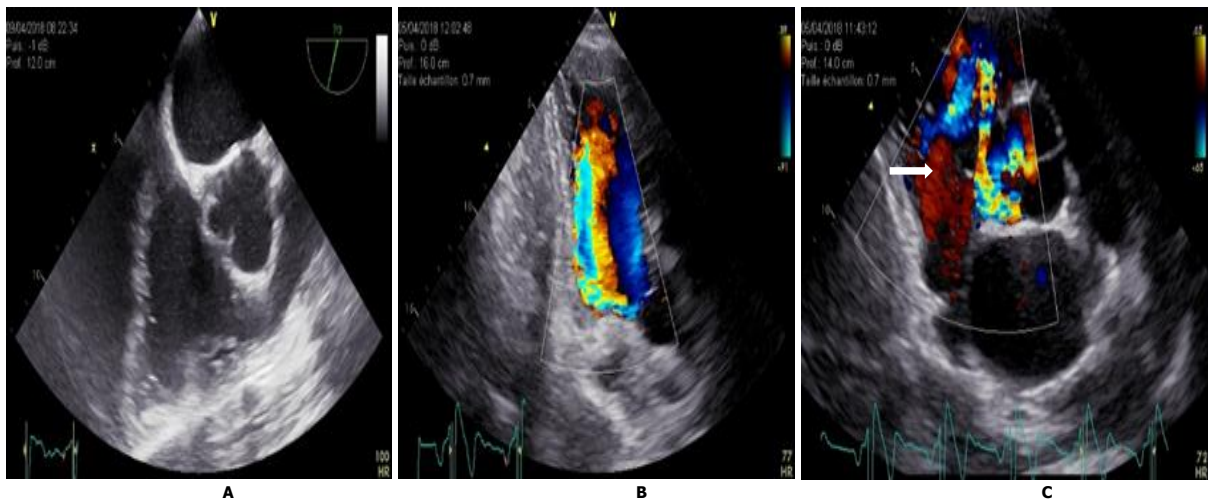


Figure 1
A: Transthoracic aspect of the right coronary
B: Transesophageal echocardiography color doppler shows severe eccentric aortic regurgitation caused by prolapse of the right coronary cusp.
C: Parasternal long-axis view of a perimembranous VSD partially occluded by the prolapsed cusp. The color flow imaging demonstrates the aortic regurgitation.

The postoperative courses were uneventful. The echocardiography done on day four post-procedure showed no ventricular septal defect. The aortic regurgitation was non-significant. The coaptation height was estimated at 9 mm. the right ventricular pressure was improved at 26mmhg. The patient was discharge at the seventh day. The echocardiography follow-up after 3 months did not show major abnormalities.

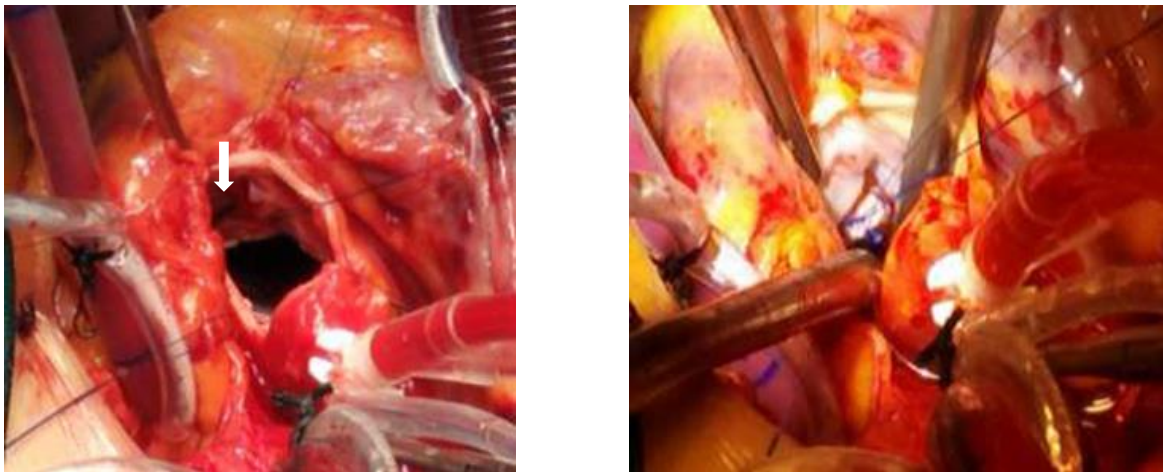


Figure 2: VSD closure with interrupted pledgeted sutures

Discussion

The Laubry-Pezzi syndrome is characterized by the association of a VSD and an aortic regurgitation. Surgery is the only valuable management option. Early diagnosis permit to treat of the VSD before the AR installment [3]. The trans-aortic approach allowed us to do a single step procedure allowing the repair of the septal and the aortic disorders at the same time. The results of surgical treatment of Laubry-Pezzi syndrome are usually satisfactory [4]. The aortic valve repair may be recommended when feasible to avoid lifetime anticoagulant therapy for such young patients [5]. After a proper assessment of the septal and valvular status; simple suturing provides enhanced recovery. The effectiveness of this technique should be always checked by an intraoperative transesophageal echocardiography.

Conflict of Interest: None

References

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