Global Academic Journal of Medical Sciences

Available online at www.gajrc.com
DOI: 10.36348/gajms.2023.v05i06.002

OPEN ACCESS

ISSN: 2706-9036 (P)
ISSN: 2707-2533 (O)

Original Research Article

Laubry-PEZZI Syndrome Surgical Repair in Adult: A Case Report

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Article History
Received: 05.07.2023
Accepted: 07.08.2023
Published: 29.11.2023

Abstract: The presence of aortic regurgitation (AR) in the setting of the ventricular septal defect (VSD) has always been a management challenge. A 21-year-old male patient presented to the Cardiovascular surgery department with a restrictive perimembranous VSD with severe aortic regurgitation (AR). VSD was first diagnosed at 3 years of age. The surgical correction was decided when he was symptomatic of severe aortic regurgitation. Trans-aortic approach was performed to close the ventricular septal defect and correct the aortic regurgitation simultaneously. Surgical management was followed by a complete heart block.

Keywords: Laubry PEZZI syndrome, Ventricular septal defect, Aortic regurgitation, Surgical repair, Complete heart block.

INTRODUCTION

Aortic regurgitation resulting from suction of right coronary or non-coronary cusp by Venturi effect in the VSD (ventricular septal defect) is associated with Laubry Pezzi syndrome. It is most commonly found in infundibular ventricle septal defect but can also be encountered with perimembranous ventricle septal defect [1]. Early ventricle septal defect closure is the main action to prevent the onset of aortic regurgitation. The management of this rare pathology is still non-consensual regarding operative timing and technique [2]. Laubry PEZZI syndrome with moderate or severe aortic regurgitation represent a challenging surgical issue. In the treatment of aortic regurgitation, aortic valve repair was an appealing alternative to aortic valve replacement. Permanent complete heart block (CHB) is a significant intracardiac repair complication for congenital heart disease [3].

CASE REPORT

A 21year-old man patient with New York Heart Association (NYHA) functional dyspnea class III symptoms was admitted in our cardiovascular surgery department with Laubry PEZZI syndrome as diagnosis. The patient had been followed since the age of 3 months for restrictive perimembranous ventricular septal defect. The clinical examination noted a diastolic murmura on aortic focus. Preoperative trans thoracic echocardiography (TEE) revealed severe aortic regurgitation caused by prolapse of the right coronary cusp in the restrictive perimembranous ventricular septal defect (6mm of diameter), moderately dilated left ventricle with preserved left ventricle function (left ventricular ejection fraction=58%). Surgery management was indicated. Under general anesthesia, a transoesophageal echocardiography probe was inserted before surgery and a median sternotomy incision was performed. Conventional cardiopulmonary bypass was established between bicaval venous canulation and aortic one under moderate hypothermia. After aortic cross-clamping, the ascending aorta was opened through an S incision allowing easy trans-aortic access and direct blood

Citation: Nijimbere Macédoine, Mpira Marien Yves, Son Moussa, Thiombiano Abdoulaziz, Arafaou Dicko, Zalikatou Abdoulkrim, Boumzebra Drissi (2023). Laubry-PEZZI Syndrome Surgical Repair in Adult: A Case Report. *Glob Acad J Med Sci*; Vol-5, Iss-6 pp- 277-279.

cardioplegia administration in the coronary ostia that was initially delivered through an aortic root cannula.

The closure of perimembranous ventricular septal defect was performed through trans-aortic exposure using three interrupted suture stitches. A triple aortic commissuroplasty with pledged suture stitches and a plication of the free edge of the right coronary cusp were achieved to restore normal height of right coronary cusp and aortic valve anatomy, hence a normal surface of cusps coaptation. The procedure control with transoesophageal echocardiography was performed intraoperatively and showed a minimal aortic regurgitation. Cardiac activity resumed in Complete cardiac block requiring the temporary pacemaker placement before the cardiopulmonary bypass stop. The echocardiography done on day seven post-procedure showed no residual ventricular septal defect, non stenosing plasty and a minimal aortic regurgitation. Despite the corticotherapy, the complete cardiac block was still permanent and on day 23, the implantation of permanent pace maker was performed and the patient was discharge at the twenty fourth day.

DISCUSSION

Laubry-Pezzi syndrome is a congenital heart disease and is still no agreement about its surgical management [4]. Sub-arterial ventricular septal defect are complicated by aortic regurgitation five times more than perimembranous ventricular septal defect [5]. Addressing the aortic valve at an earlier age has been shown to provide better outcomes [6]. If left untreated, aortic regurgitation will progress to a more severe form in the older age, necessitating aortic valve replacement rather than aortic valve repair and it has been pointed out that the grade of aortic regurgitation is associated with the size of ventricular septal defect [7]. The delayed onset of severe aortic insufficiency extending over 21 years in our patient would be justified by the ventricular septal defect anatomical location at the membranous septum despite its small size which could accelerate its severity. The trans-aortic approach allows a single-step procedure to correct the ventricular septal defect and the aortic regurgitation simultaneously [2]. Our case adheres to this rule. The VSD was directly closed using three interrupted sutures stretches through trans-aortic approach. This approach ensured effective closure and allowed restoration of the normal blood flow pattern between the ventricles.

Owing to the prolapse of the right coronary cusp into the VSD, the anatomy of the aortic valve was affected, resulting in aortic regurgitation. A triple aortic commissuroplasty was carried out, which involved pledgeted suture stitches to repair and reinstate the normal height of the right coronary cusp and the aortic valve anatomy. This procedure aimed to ensure proper coaptation of the aortic valve cusps, thereby minimizing aortic regurgitation. The echocardiography control revealed minimal residual regurgitation, sparing the patient from the need for surgical revision. In Laubry PEZZI syndrome, the initial lesion is a ventricular septal defect. Aortic regurgitation progressively worsens over time depending on the location and size of the ventricular septal defect. Perimembranous ventricular septal defects involving the inlet are more susceptible to cardiac heart block after repair [8]. This vulnerability is due to the close relationship between the HIS bundle and the posterior and inferior margins of the ventricular septal defect, making it more prone to operative traumatism; the margin of error can be smaller [9]. Rare instances of complete cardiac heart block following the closure of small perimembranous VSD have been reported by Andersen *et al.* [10], and this was the situation with our patient. In this case, the patient necessitated the insertion of a temporary pacemaker before discontinuing cardiopulmonary bypass. Regrettably, despite corticotherapy, the cardiac block persisted permanently, necessitating the placement of a permanent pacemaker to regulate the heart's rhythm. The implantation of a dual chamber permanent pacemaker took place on D 23, and the patient was discharged from the hospital on D 24.

CONCLUSION

Surgical treatment of Laubry-Pezzi syndrome through a trans-aortic approach can effectively repair both the septal defect and the aortic disorder simultaneously. Early detection, follow-up, and timely surgical management are essential in reducing the need for aortic valve replacement. However, complete heart block, though rare, can occur as a complication, necessitating the implantation of a permanent pacemaker.

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