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# Infective Endocarditis Revealing Laubry Pezzi Syndrome: A Rare Case Report

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***Authors' contributions***

*This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.*

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***Case Study***

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## ABSTRACT

**Background:** Infective endocarditis is still a frequent, life-threatening condition, despite advances in diagnosis and treatment. It can occur in a healthy heart, but more often in a pathological heart, as the congenital heart disease without surgery, or with a residual shunt or prosthetic material.
**Case Report:** We report the case of a 24-year-old patient with no particular pathological antecedents, presenting with prolonged fever, asthenia and altered general condition. Cardiac investigation revealed infective endocarditis in Laubry-Pezzi syndrome with perimembranous ventricular septal defect (VSD) and severe aortic leakage. Management consisted of effective antibiotic therapy combined with aortic valve replacement and VSD closure. The outcome was favorable.

*Keywords: Endocarditis; Laubry-Pezzi syndrome; antibiotic therapy.*

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## 1. INTRODUCTION

The Laubry-Pezzi syndrome is a rare congenital heart disease first described in 1921, associating an interventricular communication, usually perimembranous, with an aortic leak secondary to partial closure of the communication by valvular tissue. In addition to heart failure, the risk of this condition is infective endocarditis, which can be a revelation of the disease, as in our patient's case.

## 2. CASE REPORT

We report the case of a 24-year-old patient, with no particular pathological history, who presents for a prolonged fever for 3 weeks, NYHA stage III dyspnea, asthenia and deterioration in general health.

On admission, the patient was pale, conscious, hemodynamically and respiratory stable, febrile at 38.1. Cardiovascular auscultation found a diastolic murmur at the aortic focus, associated with a left lateral sternal murmur. The rest of the examination found splenomegaly, purpura in both lower and a bad oral state.

Biology showed hypochromic microcytic inflammatory anemia at 8.6 g/dL with a serum ferritin level of 245 ng/ml, neutrophils at 11,850/µL and thrombocytosis at 520,000/µL, elevated CRP at 121 mg/ dL, an elevated PCT at 1.09 mg/l, and an elevated rheumatoid factor at 203.4 IU/ml. Blood cultures isolated group A hemolytic streptococcus.

The performed transthoracic echocardiography found two vegetations of 11x26 mm and 18x9 mm on the edges of a restrictive perimembranous interventricular communication of 3 mm closed partially by tissue of the aortic valve, causing a severe aortic leak, forming a Laubry-Pezzi syndrome, moreover the left ventricle was dilated, with a blade of pericardial effusion.

Thoraco-abdominal computed tomography (CT) revealed homogeneous splenomegaly. Cerebral CT, sinus X-ray and fundus were without abnormalities. The treatment was based on the administration of vancomycin (30 mg/kg/day IV divided into two doses) for 4 weeks and gentamycin (4 mg/kg/day IV as a single dose) for 2 weeks. The evolution was favorable with disappearance of the fever, normalization of the biological assessment. The operative indication for replacement of the aortic valve with closure of the interventricular communication was raised.

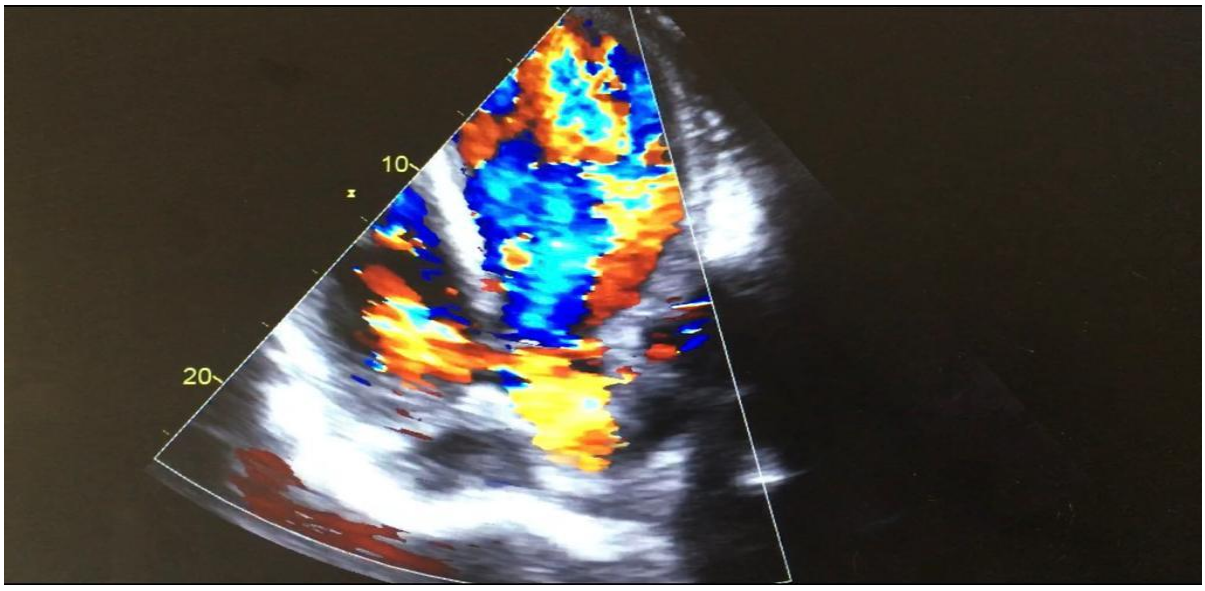


**Fig. 1. Apical 5-chamber echocardiographic section with color Doppler showing restrictive perimembranous interventricular septal defect associated with severe aortic leak forming Laubry- Pezzi syndrome**

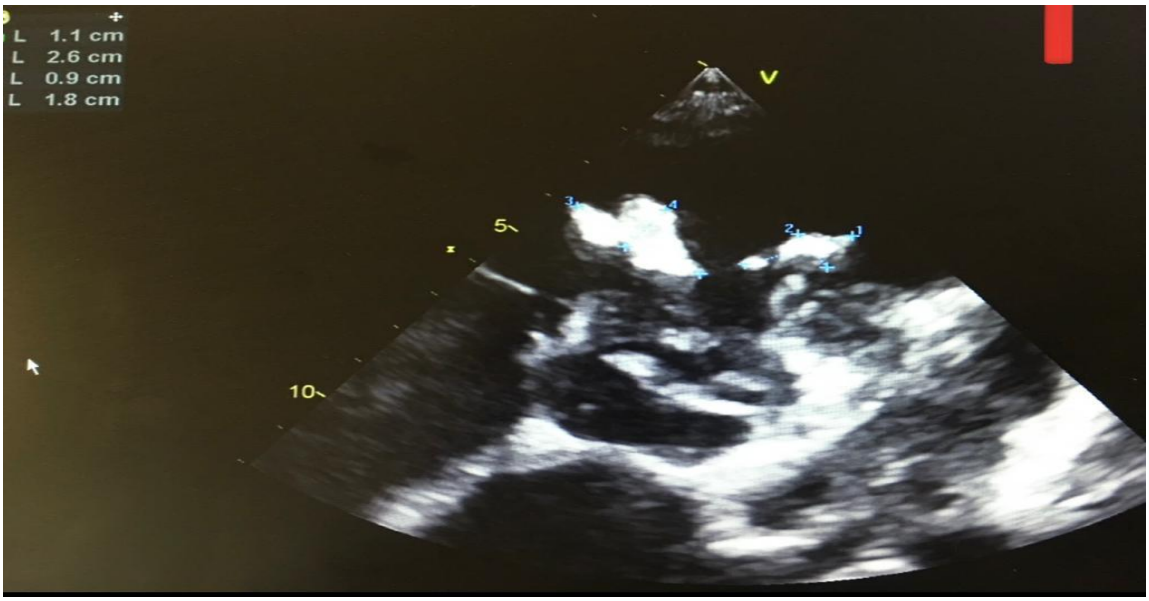


**Fig. 2. Short axis parasternal echocardiographic section showing two vegetations at the edges of a perimembranous interventricular communication**

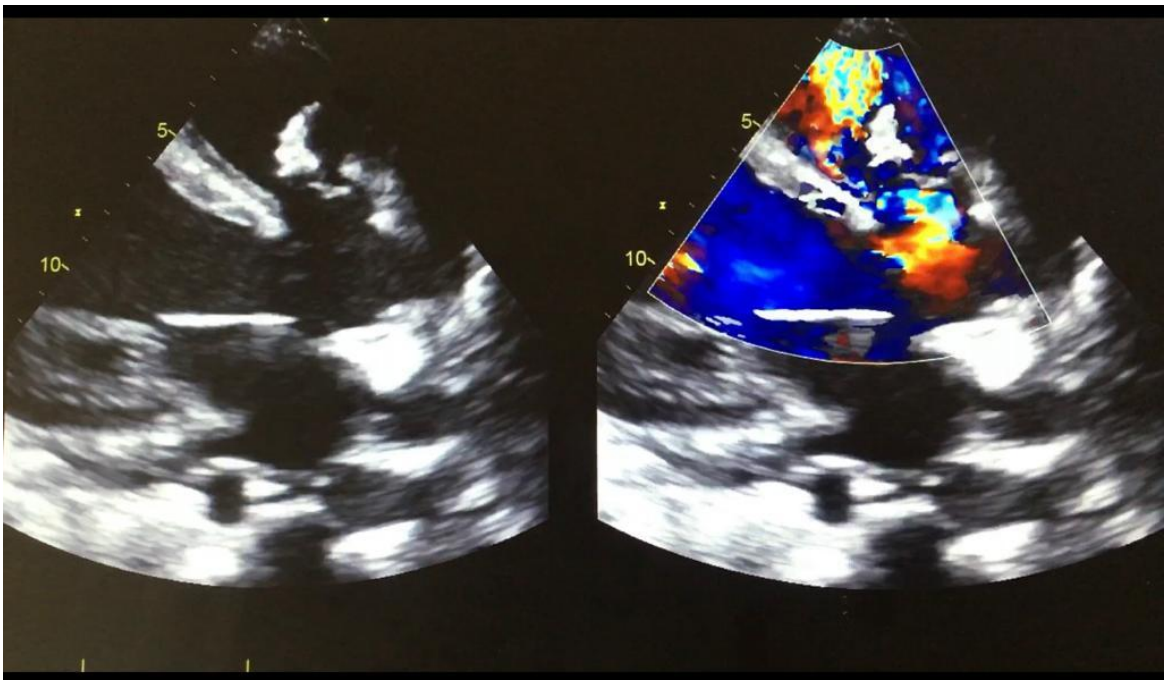


**Fig. 3. Long axis parasternal echocardiographic section showing perimembranous interventricular communication with vegetation inserted on its edges**

## 3. DISCUSSION

The incidence of infective endocarditis is estimated at around 30 cases per million inhabitants per year in general population studies conducted in Western countries. Despite advances in diagnosis and treatment, it has an unfavorable prognosis, with a mortality rate of approximately 20% [1]. Although it can occur in a healthy heart, it is most often associated with an underlying cardiac pathology [2]. In developing countries, infective endocarditis related to rheumatic valve disease is the most common [3], but the profile of patients with infective endocarditis is changing in developed countries. Endocarditis due to rheumatic or degenerative

valvular disease is becoming less frequent, in favor of endocarditis due to congenital heart disease (CC) [4]. Bacterial transplants can affect "corrected" congenital heart disease, not operated on or having benefited from palliation. It is assumed that surgical repair decreases or even eliminates the risk if there is no residual injury. The repair of complex heart disease, which involves the placement of prosthetic valves or prosthetic tubes, creates sites at risk of infection, or leaves valve lesions or other shunts, presenting a lower risk, but not negligible [5].

Laubry-Pezzi syndrome is a rare congenital heart disease [6,7]. The prevalence of aortic valve prolapse in perimembranous ventricular septal defects (VSDs) is estimated to be around 5-8%, with a higher occurrence in males [8,9]. The first clinical description of Laubry-Pezzi syndrome was documented in 1921 by Charles Laubry and Cesare Pezzi, based on a patient presenting with VSD and aortic regurgitation caused by the prolapse of an aortic cusp [6,10]. This syndrome may eventually lead to dilatation of the aortic sinus related to the affected leaflet, resulting from a discontinuity between the media and the aortic annulus. Anatomically, this association seems to be more common in perimembranous VSDs, leading to the prolapse of the right coronary cusp or, less frequently, the non-coronary cusp due to a discontinuity between the ventricular septum and the aortic sinus [10,11]. The pathophysiology of the syndrome is explained by the Venturi effect, where the restrictive flow through the VSD creates a low-pressure zone that pulls in the adjacent cusp, resulting in aortic prolapse and regurgitation [12]. The diagnosis primarily relies on transthoracic echocardiography (TTE) and should be performed before the emergence of aortic regurgitation.

The management of this rare syndrome lacks consensus regarding the optimal timing and surgical approach [13]. However, early closure of the VSD is recommended to prevent the onset or progression of aortic insufficiency [14,15]. In cases where aortic valve prolapse and regurgitation have already occurred, VSD closure alone may not be sufficient, necessitating the need for aortic valve repair or replacement. Therefore, early closure of the VSD is crucial. Aortic valve repair is preferable in young patients to avoid the need for lifelong anticoagulation [16].

Patients with Laubry-Pezzi syndrome are at an increased risk of endocarditis as the aortic valve dysfunction progresses [14]. Ventricular septal defect is the second most common congenital heart disease after bicuspid aortic valve, but it is the first in terms of infective endocarditis. Various studies and registries have shown that among congenital heart disease endocarditis, VSD is the most common, with patients with VSD having six times the risk of developing infective endocarditis than the general population [17,18]. The clinical presentation of infective endocarditis is highly variable, but fever is almost always present and is the main revealing symptom. Sometimes severe heart failure can be a telltale sign or complicate the course of the disease.

The use of echocardiography, whether transthoracic echocardiography (TTE) or transesophageal echocardiography (TEE), is of critical importance in the management and follow-up of infective endocarditis (IE). It is essential to perform this examination as soon as IE is suspected. TEE is recommended in cases where the patient is known to have a prosthetic valve or intracardiac device, if the TTE is negative but the clinical suspicion of IE is high, especially if the TTE is not optimal. However, ultrasound diagnosis can be difficult in complex heart disease with valvular abnormalities and multiple shunts: nearly 65% of infective endocarditis in these complex heart diseases do not show obvious echocardiographic lesions. Indeed, it may sometimes be impossible to distinguish infectious lesions from pre-existing abnormalities, or these may be invisible if they are located on palliative systemic-pulmonary shunts [5]. Generally, these difficulties do not arise in the case of infective endocarditis on ventricular septal defect [17].

## 4. CONCLUSION

Laubry-Pezzi syndrome is a rare congenital heart disease, associating interventricular communication with aortic insufficiency of variable severity. It represents a favorable ground for infective endocarditis, which can be indicative of this pathology, as is the case of our patient. Once the diagnosis has been made, surgical management is indicated, with closure of the interventricular communication and aortic replacement or repair, depending on the lesions and the severity of the aortic insufficiency.

## CONSENT

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

## ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

## COMPETING INTERESTS

Authors have declared that no competing interests exist.

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