

INFECTIVE ENDOCARDITIS REVEALING LAUBRY PEZZI SYNDROME: A RARE CASE REPORT

ABSTRACT:

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.

We report the case of a 24-year-old patient with no particular pathological antecedents, presenting with prolonged fever, associated with asthenia and altered general condition. Cardiac investigation revealed infective endocarditis in Laubry-Pezzi syndrome with perimembranous ventricular septal defect and severe aortic leakage. Management consisted of effective antibiotic therapy combined with aortic valve replacement and IVC closure. The outcome was favorable.

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.

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 associated with 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 at the radius of the wheel. 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/ μ L and thrombocytosis at 520,000/ μ L, 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.

Comment [u1]: Try to make it clear and include recent findings, then merge them into one paragraph.

Comment [u2]: Try to include key words and make it in alphabetical order.

Comment [u3]: Try to include a statement of the problems, general and specific objectives, a hypothesis (a research question), and the scope of the research.

Comment [u4]: Try to include a research period, inclusion and exclusion criteria, data quality checking issues, and the method of data analysis.

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.

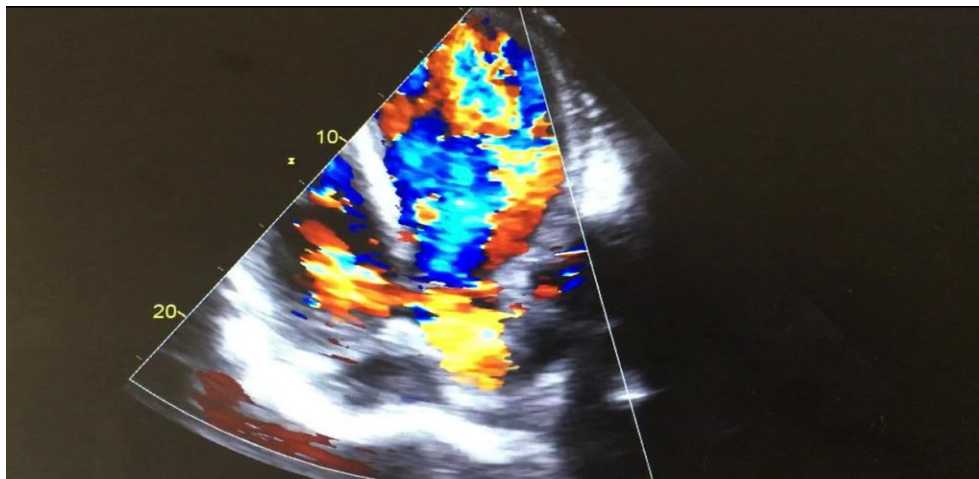


figure 1: apical 5-chamber echocardiographic section with color Doppler showing restrictive perimembranous interventricular septal defect associated with severe aortic leak forming Laubry Pezzi syndrome

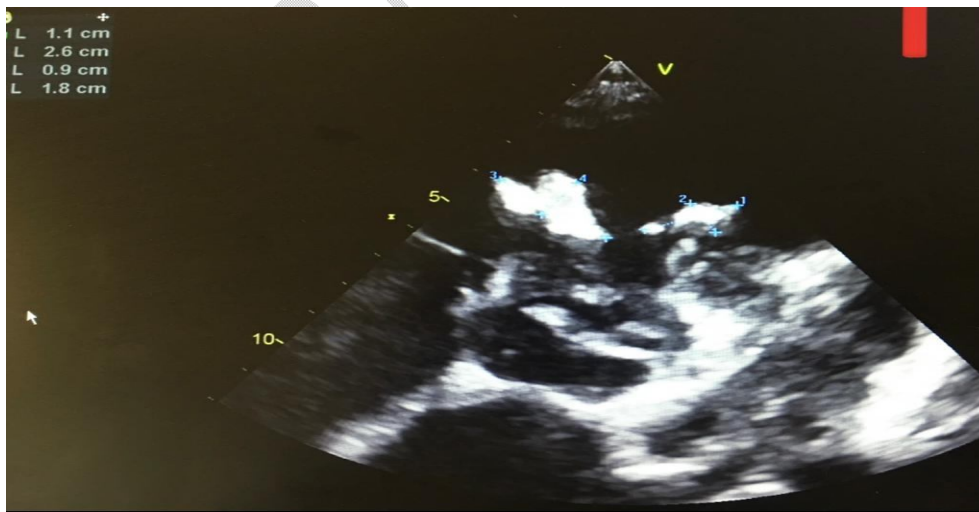


figure 2 : short axis parasternal echocardiographic section showing two vegetations at the edges of a perimembranous interventricular communication

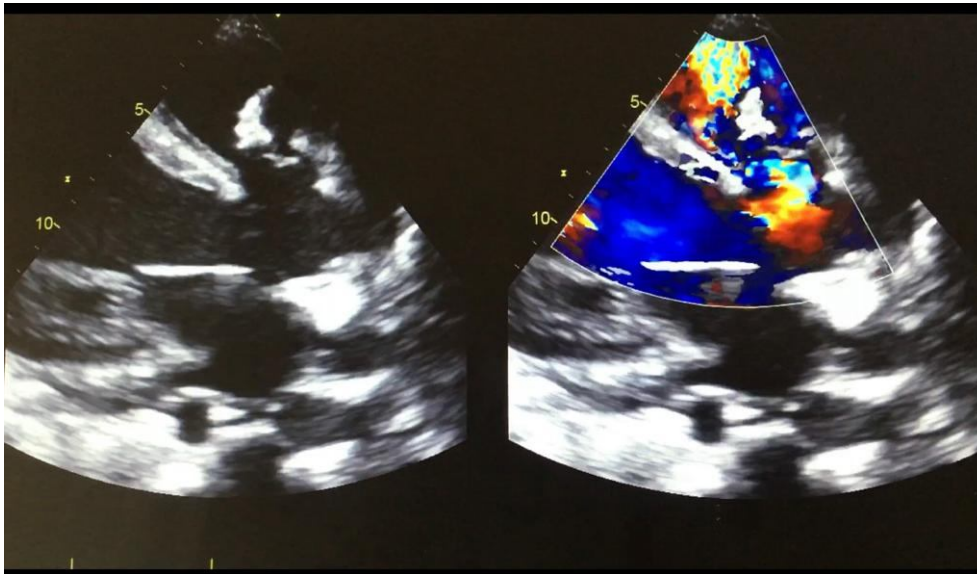


Figure 3 : long axis parasternal echocardiographic section showing perimembranous interventricular communication with vegetation inserted on its edges

Comment [u5]: Try to modify the case reporting format to be sound for the readers.

DISCUSSION:

Infectious endocarditis is a rare disease with an annual incidence of approximately 10 cases per 100,000 inhabitants. 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 (VSD) 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 [18-17]. 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 ETT is negative but the clinical suspicion of IE is high, especially if the ETT 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 (VSD)[17].

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, depending on the lesions and the severity of the aortic insufficiency, aortic replacement or repair.

REFERENCES :

- 1-Fabio Chirillo, Pompilio Faggiano, Moreno Cecconi, Antonella Moreo, Angelo Squeri, Oscar Gaddi, Enrico Cecchi. Predisposing cardiac conditions, interventional procedures, and antibiotic prophylaxis among patients with infective endocarditis. Am Heart J. September 2016; Volume 179: 42-50.
- 2-Gilbert Habib, Patrizio Lancellotti, Manuel J Antunes, Maria Grazia Bongiorno, Jean-Paul Casalta, Francesco Del Zotti, Raluca Dulgheru, Gebrine El Khoury, Paola Anna Erba, Bernard Jung, Jose M Miro, Barbara J Mulder, Edyta Plonska Gosciniak, Susanna Price, Jolien Roos-Hesselink, Ulrika Snygg Martin, Franck Thuny, Pilar Tornos Mas, Isidre Vilacosta, Jose Luis Zamorano. 2015

Comment [u6]: Try to include a recent citation in line with the issue.

Comment [u7]: Try to include the recommendation based on the research review output.

- ESC Guidelines for the management of infective endocarditis. *Rev Esp Cardiol*. 2016;69:69 - Vol. 69 Num.01 DOI: 10.1016/j.rec.2015.100.
3. Maharaj Breminand, Parrish Andrew. Prevention of infective endocarditis in developing countries. *Cardiovasc J Afr*. 2012 Jul; 23(6):303-5.
 4. Jae Eun Baek, Su Jin Park, Saet Byul Woo, Jae Young Choi, Jo Won Jung, Nam Kyun Kim. Changes in patient characteristics of infective endocarditis with congenital heart disease: 25 years experience in a single institution. *Korean Circ J*. 2014 Jan;44(1):37-41.
 5. Di Filippo S, Sassolas F, Celard M, Ducreux C, Henaine R, Ninet J, Bozio A. Infective endocarditis in children. *Arch Pediatr*. 2006 Jun; Volume 13 (6): pages 629-630
 - 6- Laubry C, Pezzi C. Treatise on congenital heart diseases. Paris: JB Bailliere; 1921. Quoted by Labury C, Routier D, Soulie P. The blows of Roger's disease. *Rev Med Paris* 1933;50:439-48.
 - 7-Abir R, Djamal K, Toufik S. Surgical repair of perimembranous ventricular septal defect and aortic regurgitation in an adult patient with Laubry-Pezzi syndrome. *Jr Med Res* 2020;3:15-7.
 - 8- Aimar F, Lakehal R, Bendjaballah S, et al. Aortic valve repair versus replacement in Laubry and Pezzi syndrome. *Arch Cardiovasc Dis Suppl* 2017;9:287-8.
 - 9-Schmaltz AA, Schaefer M, Hentrich F, et al. Ventricular septal defect and aortic insufficiency: pathophysiologic aspects and therapeutic consequences. *J Cardio* 2004;93:194-200.
 - 10- Martínez-Quintana E, Rodríguez-González F, López-Gude MJ. Laubry-Pezzi syndrome with aortic root dilatation treated with a Bentall and De Bono procedure. *World J Pediatr Congenit Heart Surg* 2013;4:299-301.
 - 11- Pontailier M, Gaudin R, de Bellaing AM, et al. Surgical repair of concomitant ventricular septal defect and aortic cusp prolapse or aortic regurgitation, also known as the Laubry-Pezzi syndrome. *Ann Cardiothorac Surg* 2019;8:438-40.
 - 12- Schmaltz AA, Schaefer M, Hentrich F, et al. Ventricular septal defect and aortic insufficiency: pathophysiologic aspects and therapeutic consequences. *J Cardio* 2004;93:194-200.
 - 13- Jung H, Cho JY, Lee Y. Progression of aortic regurgitation after subarterial ventricular septal defect repair: optimal timing of the operation. *Pediatr Cardiol* 2019;40:1696-702.
 - 14- Piazza F, Santoro G, Russo MG. Aortic insufficiency due to ventricular septal defect (Laubry-Pezzi syndrome). *J Cardiovasc Med* 2013;14:164-5.
 - 15- Krishnasamy S, Sivalingam S, Dillon J, et al. Syndrome of ventricular septal defect and aortic regurgitation – a 22-year review of its management. *Braz J Cardiovasc Surg* 2021;36:807-16.
 - 16- Yacoub MH, Khan H, Stavri G, et al. Anatomic correction of the syndrome of prolapsing right coronary aortic cusp, dilatation of the sinus of Valsalva, and ventricular septal defect. *J Thorac Cardiovasc Surg* 1997;113:253-61.
 17. Baumgartner H, Bonhoeffer P, De Groot NM, de Haan F, Deanfield JE, Galie N, Gatzoulis MA, Gohlke-Baerwolf C, Kaemmerer H, Kilner P, Meijboom F, Mulder BJ, Oechslin E, Oliver JM, Serraf A, Szatmari A, Thaulow E, Vouhe PR, Walma E; Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC); Association for European Paediatric Cardiology (AEPC); ESC Committee for Practice Guidelines (CPG). ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). *Eur Heart J*. 2010 Dec;31(23):2915-57.
 18. Loureiro-Amigo J, Fernández-Hidalgo N, Pijuan-Domènech A, Dos-Subirà L, Subirana Domènech T, González-Alujas T, González-López JJ, Tornos-Mas P, García-Dorado D, Almirante B. Infective endocarditis in adult patients with congenital heart disease: experience from a reference centre. *Enferm Infecc Microbiol Clin*. 2016 Feb 6. PubMed | Google Scholar