

CASE REPORT

Perforation of the non-coronary leaflet in a patient with Laubry-Pezzi Syndrome

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Laubry-Pezzi syndrome is a rare cause of aortic insufficiency. It is characterized by ventricular septal defect and aortic valve prolapse. We report the case of a 15-year-old patient with Laubry-Pezzi syndrome. During the surgical procedure it is noted perforation of the non-coronary leaflet secondary to Venturi effect.

Key words: Aortic valve prolapse; Aortic valve regurgitation; Congenital heart disease; Laubry-Pezzi syndrome; Ventricular septal defect.

El síndrome de Laubry-Pezzi es una causa rara de insuficiencia aórtica. Se caracteriza por comunicación interventricular y prolapso de la válvula aórtica. Presentamos el caso de un paciente de 15 años de edad con síndrome de Laubry-Pezzi. Durante el acto quirúrgico se observó una perforación de la valva no coronaria aórtica secundaria a efecto Venturi.

Palabras clave: Prolapso valvular aórtico; Insuficiencia aórtica; Cardiopatía congénita; Síndrome de Laubry-Pezzi; Comunicación interventricular.

Cir Card Mex 2023; 8(3): 78-79.

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Laubry-Pezzi syndrome is a rare, non-syndromic cardiac malformation characterized by an interventricular septal defect (VSD) and aortic insufficiency, which produce prolapse of the right coronary leaflet, and less frequently of the non-coronary leaflet [1]. The prolapse is produced by Venturi effect [2]. It is a rare cause of aortic insufficiency and was described in 1921 [3]. However, as it is a rare pathology, both the time for surgery and surgical technique are still a matter of debate [3].

CLINICAL CASE

We present the case of a 15-year-old patient with a heart murmur in preschool age, diagnosed with a subaortic ventricular septal defect with aortic valve prolapse and moderate aortic insufficiency. In the subsequent evaluations, he presented an impairment in functional class. Echocardiography showed an increase in aortic insufficiency. It was decided to close the defect.

Operation was performed by using median sternotomy and cardiopulmonary bypass. Bicaval cannulation and retrograde cardioplegia were used. The subaortic interventricular defect was approached by means of right atriotomy (**Fig. 1**). A circular

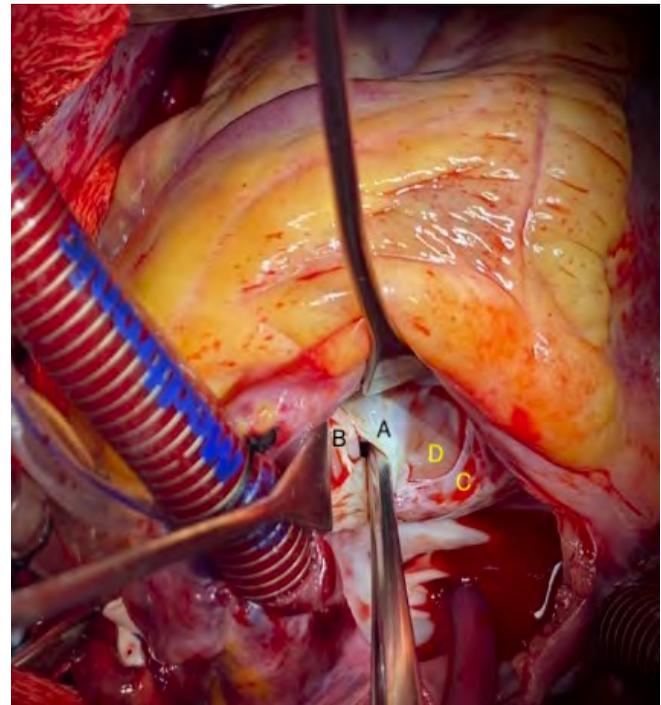


Figure 1. Intraoperative image. Transtricuspid view. The Interventricular communication (A), non-coronary valve (B), Tricuspid Valve (C), Interventricular septum (D) is observed.

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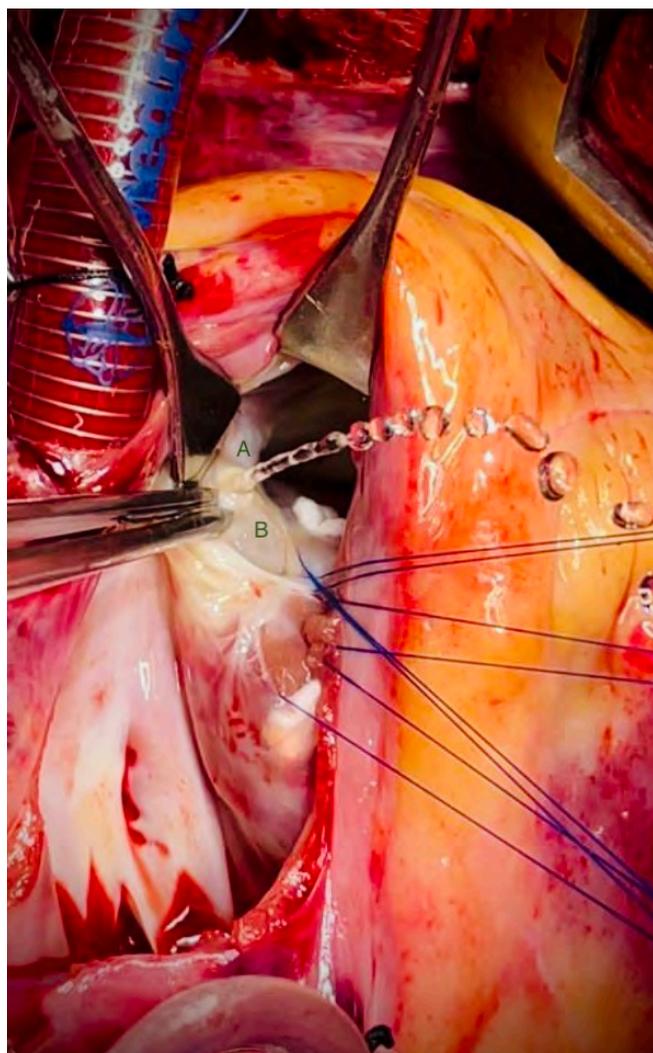


Figure 2. Intraoperative image. Transtricuspid view. Perforation of the non-coronary valve (A) with prolapse and hydraulic test (B).

perforation is also observed in the free edge of the non-coronary aortic valve, which included approximately a heavy fibrous edge of 0.5 cm (Fig. 2). The defect was closed. Cardiopulmonary bypass time and aortic cross-clamping time were of 151 min and 133 min, respectively. The intraoperative echocardiogram shows adequate ventricular function with a trileaflet aortic valve with trivial aortic valve regurgitation. In the immediate postoperative period upon admission to intensive care unit, no mechanical ventilation was required, with only minimal inotropic support for 36 hours. Postoperative course was free of complications.

COMMENT

The association of infundibular VSD with prolapse and aortic insufficiency is rare with an approximate incidence of 5 to 10%, being the prolapse progressive and with the presence of small defects it has been observed that aortic insufficiency tends to evolve more rapidly [4].

Being a rare pathology, the decision of surgical intervention is a controversial issue. However, some authors refer that it should be considered like any other interventricular defect; that is, surgical closure should be performed when aortic insufficiency is observed regardless of the hemodynamic repercussion of heart disease, since its development can complicate the patient's prognosis and survival [4]. Surgical results are usually satisfactory.

FUNDING: None

DISCLOSURE: The authors have no conflicts of interest to disclose.

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