

## Isolated Infundibular Pulmonary Stenosis with Infective Endocarditis: a Rare Ecocardiographic Finding

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### Introduction

The right ventricle (RV) consists of three portions: the inflow tract, trabecular region, and outflow tract, also called infundibulum, which separates the tricuspid and pulmonary valves. Some congenital heart diseases include abnormalities in this area and can cause obstruction of the right ventricular outflow tract (RVOT). Infundibular pulmonary stenosis (IPS) is one of these cardiopathies and is often associated with other abnormalities, especially interventricular septal defect but, as an isolated presentation, it is considered a rare finding. Heart diseases may result in serious complications such as heart failure and infective endocarditis (IE). The latter has vegetation as a typical lesion. Vegetation is a mass of platelets and fibrin, in which microorganisms and inflammatory cells are captured. It can be precipitated by a valvulopathy or endocardial lesion. This report describes the case of a young patient admitted to a tertiary hospital with rare congenital pathology, whose diagnosis was made as a result of infective endocarditis.

### Case report

A 20-year-old female patient was admitted with fatigue and intermittent fever associated with systemic symptoms, such as chills, asthenia and anorexia for 30 days. The patient had significant worsening of tiredness, dyspnea and edema of lower limbs, as well as episodes of cyanosis. She denied the use of medication and illicit drugs. She also reported dental extraction in the past six months.

At admission, the patient was conscious and oriented, with tachydispnea, hypoxemic (89% oxygen saturation), tachycardic, normotensive, with pathological jugular stasis and lower limb edema ++/4+. At the cardiovascular examination, the pulses were symmetrical and there was systolic tremor in the precordium. On cardiac auscultation, cardiac rhythm was regular with two sounds, with systolic murmur in pulmonary focus 5+/6+. Pulmonary auscultation revealed crackling rales at the bases of both hemithorax. Abdominal palpation showed hepatomegaly.

### Keywords

Stenosis, Pulmonary; Ultrasonography; Endocarditis.

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Electrocardiogram revealed overload of right chambers with strain pattern. Chest radiography showed cardiomegaly at the expense of the right chambers, interstitial infiltrate at the bases of both hemithorax and kyphoscoliosis.

Transthoracic echocardiography (TTE) with color flow Doppler showed marked RV hypertrophy (Figure 1) and marked IPS (maximum pressure gradient between the RV and the distal portion of the RVOT was 162 mmHg). RV systolic function was preserved and there was mild tricuspid regurgitation. A mobile and echodense mass adhered to the infundibular endocardium suggestive of vegetation was also seen (Figure 2). Interatrial septum evidenced ostium secundum atrial septal defect (ASD) with bi-directional shunt on color flow Doppler (Figure 3). No shunt was seen at the interventricular septal level.

Considering the clinical and echocardiographic findings, in addition to the laboratory tests demonstrating leukocytosis, diagnosis of infective endocarditis was established. Soon thereafter, antimicrobial therapy including culture-guided oxacillin and gentamicin showing growth of *Streptococcus viridans* was initiated. However, after 10 days of clinical treatment with no improvement of infectious profile or the symptoms resulting from heart failure, surgical treatment was recommended.

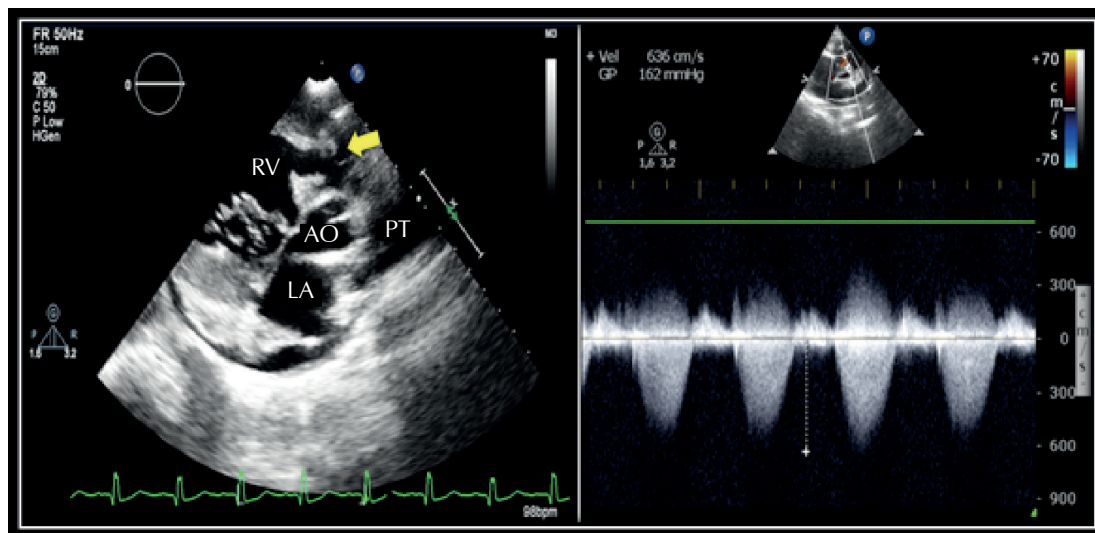
The patient was submitted to surgical intervention, which consisted of resection of the vegetation in the infundibular region, with RVOT enlargement and ASD closure. There were no adverse events in the post-operative period.

After six months of hospital discharge, the patient returned asymptomatic to the outpatient clinic. A new transthoracic echocardiogram was performed, showing persistence of right cardiac cavity dilatation, RV hypertrophy and residual infundibular pulmonary stenosis with maximum systolic gradient between the RV and the distal portion of the RVOT of 65 mmHg. No residual shunts were seen in the interatrial septum or images suggestive of vegetation (Figure 4).

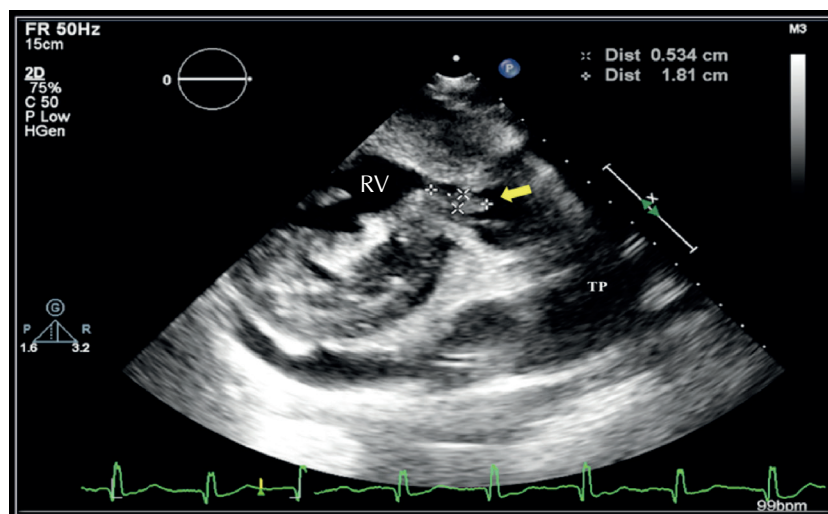
### Discussion

Isolated infundibular pulmonary stenosis is an unusual congenital anomaly.<sup>1</sup> The hemodynamic consequence of obstruction is right intracavitary right pressure and RV hypertrophy under the obstruction.<sup>2</sup> The degree of pressure rise depends on the severity of obstruction. When severe, RV systolic pressure may exceed that of the left ventricle (LV), resulting in bidirectional shunt or even flow inversion at the ASD level and, consequently, cyanosis, as in the case described here.

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**Figure 1** – Preoperative Test — Two-dimensional transthoracic echocardiography with continuous wave Doppler through short-axis parasternal window showing RV hypertrophy (left) and maximum pressure gradient between RVOT and PT of 162 mmHg (right). RA: right atrium; AO: aorta; RV: right ventricle; PT: pulmonary trunk.

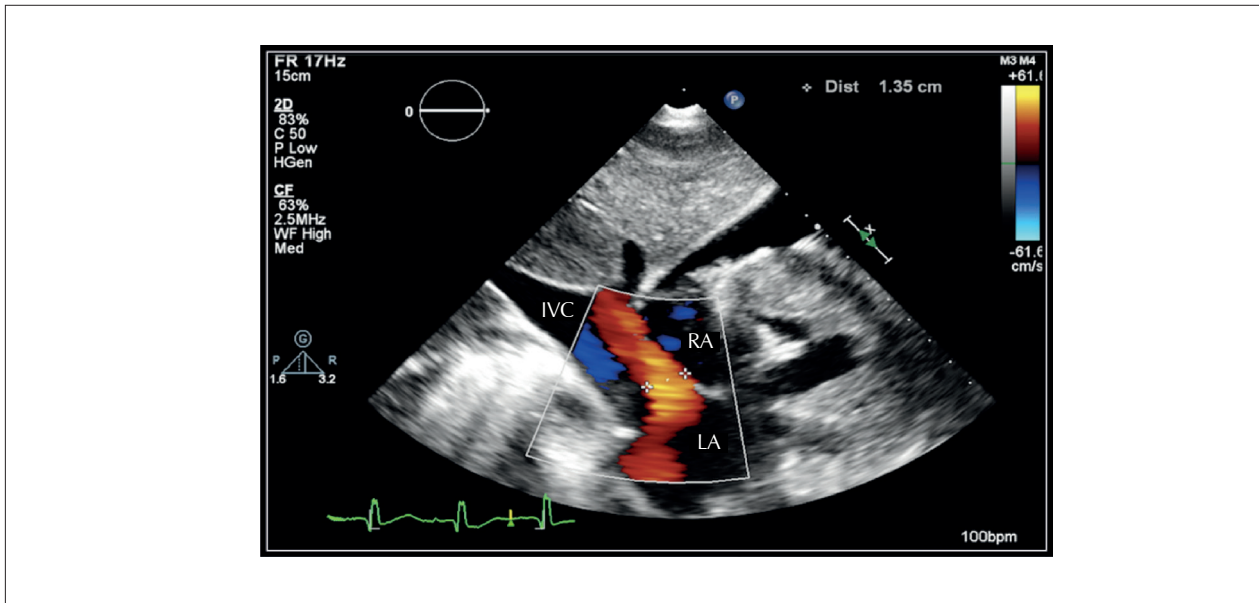


**Figure 2** – Preoperative Test — Two-dimensional transthoracic echocardiography through short axis parasternal window with image of vegetation (arrow) adhered to the infundibular narrowing area. RV: right ventricle; PT: pulmonary trunk.

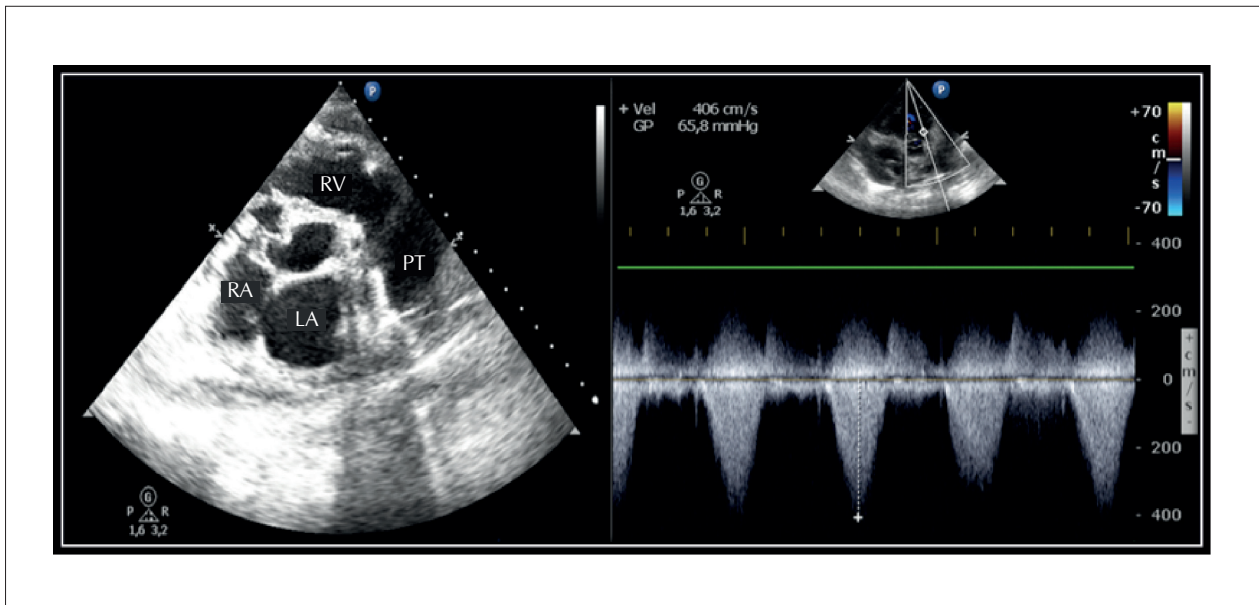
Most cases of IPS are diagnosed and treated during childhood and very few reports include cases in which the initial presentation for diagnosis and treatment occurs during adulthood.<sup>3</sup> However, the clinical presentation of isolated IPS may be silent, with no correlation between clinical manifestation and severity of RVOT obstruction. Shyu et al.<sup>4</sup> found asymptomatic adult patients with RVOT pressure gradients greater than 100 mmHg, some of them reaching 200 mmHg, which is similar to our findings.

Another peculiarity of IPS may be diagnosis through

a complication, such as IE, in the infundibular chamber, pulmonary valve and/or tricuspid valve,<sup>2</sup> as in this study. Infective endocarditis is a serious disease that, despite advances in antimicrobial treatment, remains associated with high mortality and serious complications.<sup>5,6</sup> One of the ways to reduce the incidence of this pathology in patients with congenital heart disease is adopting chemoprophylactic measures.<sup>7</sup> However, if the patient is not aware of any heart disease, prophylactic measures cannot be applied and the individual is exposed to risk of infection, as in the case of the patient described in this study.



**Figure 3** – Preoperative Test — Two-dimensional transthoracic echocardiography with color flow Doppler through subcostal window revealing atrial septal defect. LA: left atrium; RA: right atrium; IVC: inferior vena cava.



**Figure 4** – Post-operative Test — Two-dimensional transthoracic echocardiography through short axis parasternal window showing absence of vegetation in the RVOT; continuous wave Doppler revealed maximal blood pressure gradient between RVOT and PT of 65 mm Hg. RVOT: right ventricle outflow tract; PT: pulmonary trunk; LA: left atrium; RA: right atrium; RV: right ventricle.

In this case report, two-dimensional Doppler echocardiography played a fundamental role in the diagnosis of both congenital heart disease and IE, enabling rapid clinical and surgical management, thus allowing a prompt clinical, hemodynamic and infectious solution of the case, hence confirming the great importance of echocardiography, not only in the anatomic and functional diagnosis of congenital heart diseases, but also in critical and emergency situations.

The key point of IPS management is to identify when the severity of stenosis justifies the relief of obstruction. However, this situation may change in the presence of IE. With advances in surgical techniques and postoperative care systems, surgical mortality is low and the survivors' results in the medium term are satisfactory.<sup>4</sup> However, these results may also be different when there is concomitant IE. Therefore, quick diagnosis is critical to getting good results.

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### Conclusion

This report illustrates a form of congenital heart disease that is still rare in the current literature, whose diagnosis was made as a result of infective endocarditis. Detection and early intervention determined favorable prognosis.

### Contribuição dos autores

Research creation and design: Carneiro LS, Evangelista NL, Lima CJM, Diógenes MSB, Alcantara ACB, Lima LMC. Data acquisition: Carneiro LS, Evangelista NL, Lima CJM,

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