Case Report

Congenital Unprotected Tricuspid Orifice: An Adult Case Report

Orifício Tricúspide Desprotegido Congênito: um Relato de Caso em Adulto

José Thiefeson Serpa da Silva[1], Maria Suely Bezerra Diógenes[1], Nelson Lopes Evangelista[1], Ana Carolina Brito de Alcantara[2], Victoria Bianca Viana Holanda[2], Carlos José Mota de Lima[3]

De Messejana Hospital, Dr. Carlos Alberto Studart Gomes,[1] Fortaleza, CE; Christus University Center,[2] Fortaleza, CE; São Camilo Heart Center,[3] City, State, Brazil.

Introduction

The congenital unprotected tricuspid orifice (UTO) is a rare anomaly consisting of total or partial agenesis of the tricuspid valve.[1,2] This pathology represents the most extreme form of tricuspid valve dysplasia, the most common cause of isolated tricuspid regurgitation.[3] Symptom onset is more common in early childhood, consisting of congestive heart failure (CHF) and cyanosis. However, there are reports of cases in which symptom onset occurs in adulthood, consisting of severe tricuspid regurgitation and right heart failure (RHF).[2] The treatment can be surgical, consisting of total cavopulmonary anastomosis or Fontan surgery, or clinical, through symptom management with diuretics or anticoagulants, depending on the patient's needs.[3] The differential diagnoses that stand out are tricuspid valve dysplasia and Ebstein anomaly.[3]

The present report describes the clinical case of an adult patient with clinical signs of heart failure and echocardiography findings compatible with congenital UTO.

Case report

A 51-year-old man was admitted with a history of tiredness and progressive dyspnea progressing to minimal efforts, orthopnea, paroxysmal nocturnal dyspnea, and lower limb swelling. The hypothesis of Ebstein anomaly was being investigated. He reported a personal history of chronic hepatitis due to hepatitis C virus.

Upon admission, he was conscious, oriented, and normotensive with tachypnea and dyspnea, jugular vein distention at 90º, marked lower-limb edema, and a heart rate of 115 beats per minute. A cardiovascular examination revealed regular auscultation of the heart in two tachycardic times, hypophonic heart sounds without murmurs, and symmetrical pulses. Pulmonary auscultation showed crackles up to two thirds of both hemithoraxes. An abdominal examination showed a palpable and slightly painful liver 6 cm from the right costal margin.

Electrocardiography (ECG) showed atrial tachycardia, a complete right bundle branch block, low diffuse voltage in all deflections, and signs of right ventricular (RV) overload.

Transthoracic echocardiography (TTE) showed large dilation of the right heart chambers, systolic dysfunction of the RV (S', 5 cm/s), and small remnants of the anterior, septal, and posterior leaflets, with normal implantation of the tricuspid valve characteristic of congenital UTO. The inferior vena cava measured 49 mm in diameter and presented inspiratory collapse < 50%. The left ventricular ejection fraction was 58%, and the other cardiac valves showed no structural or functional changes.

Treatment with amiodarone was started to control the tachyarrhythmia and with diuretics to control the congestive symptoms. The case was discussed with the Adult Congenital Cardiopathy Team and the clinical treatment was chosen. The patient was discharged hemodynamically stable and referred for outpatient follow-up.

Discussion

The normal tricuspid orifice consists of a subvalvular apparatus and three valvular leaflets that act as a blood flow channel into the RV during diastole and prevent blood regurgitation from the RV into the right atrium (RA) during systole.[1] Without an obstructed RV outflow pathway, congenital UTO results in unrestricted communication between the RA and the RV and in right cavity dilation.[1] In adults, it presents as RHF that progresses to CHF when the left ventricle (LV) is compressed by a very dilated RV and in cases of arrhythmias. During childhood, tricuspid regurgitation is well tolerated in most cases; signs and symptoms appear only in adulthood, when significant RV dysfunction occurs.[2,4] The clinical presentation in neonates is cyanosis with CHF since the foramen ovale is patent, allowing right-to-left flow and, consequently, cyanosis. After spontaneous closure of the foramen ovale, only CHF remains.[4]

Congenital UTO is the rarest and most extreme presentation[5-7] of tricuspid valve dysplasia, with a high death rate during pregnancy, and it may have a severe clinical presentation when associated with an obstructed RV outflow pathway, such as in cyanogenic congenital heart disease like pulmonary atresia with an intact septum, in which the RV is hypoplastic.[3-7]

There are fewer than 50 cases of congenital UTO described in the literature.[7] The clinical history varies according to degree of right chamber impairment over time, and the patient may reach adulthood or older.[7] UTO must be differentiated from Ebstein malformation, since there is also leaflet dysplasia, but Ebstein cases also show a lower septal leaflet displacement of

Keywords

Congenital abnormalities; Heart diseases; Tricuspid valve.

Mailing Address: José Thiefeson Serpa da Silva •
Av. Frei Cirilo, 3480 - Messejana, Fortaleza – CE. Brazil.
E-mail: thiefeson@hotmail.com
Manuscript received 2/16/2020; revised 3/19/2020; accepted 3/31/2020

DOI: 10.5935/2318-8219.20200044

8 mm/m² or more on echocardiography unlike in UTO, in which the tricuspid leaflets are small and not inferiorly displaced.[8]

A series of patient cases described by Mohan et al.[9] showed that five of the seven analyzed adults had signs of CHF like the patient in this report, who presented with jugular vein distention, painful hepatomegaly, and lower-limb edema.

Due to a quite variable natural history, some descriptions of congenital UTO cases, such as those of Ozkutlu et al. and Mohan et al.,[5,9] corroborate this present report, in which the adult patient admitted with CHF is diagnosed only after echocardiography. Thus, the disease is only diagnosed in adulthood because the symptoms are well tolerated in patients with UTO and moderate RV dysfunction, allowing them to reach an older age.[7]

Regarding findings expected in complementary exams, the signs of right chamber overload and right branch block on the ECG stand out as described by Gupta et al.[4] In the case described here, there were signs of RV overload and complete right branch block, in addition to signs of low diffuse voltage in deflections in most leads (Figure 1), a common finding in hearts with large chamber dilation. In addition, the TTE performed on the patient in the present study confirmed the diagnosis (Figures 2–4), as he had been admitted with suspected Ebstein anomaly, the main pathology in the differential diagnosis of congenital UTO. Furthermore, it presented a large dilation of the right cavities and contractile RV dysfunction, showing the importance of this complementary exam for assessing right chamber involvement severity, mainly the RV function, and excluding other causes of right chamber dilation and tricuspid valve malformation. Cardiac catheterization may show increased atrial pressure, a prominent V wave, and the presence of interatrial shunt if there is a patent foramen ovale or interatrial communication.[4,5] It was not necessary to perform cardiac catheterization in the patient reported here once conservative treatment was indicated by the adult congenital heart disease team.

In addition to Ebstein anomaly, the malformation first investigated in the patient before hospital admission, tricuspid valve dysplasia and Uhl anomaly are part of the differential diagnosis of congenital UTO, the congenital causes of tricuspid regurgitation with marked RA and RV dilation.[1,3] However, this

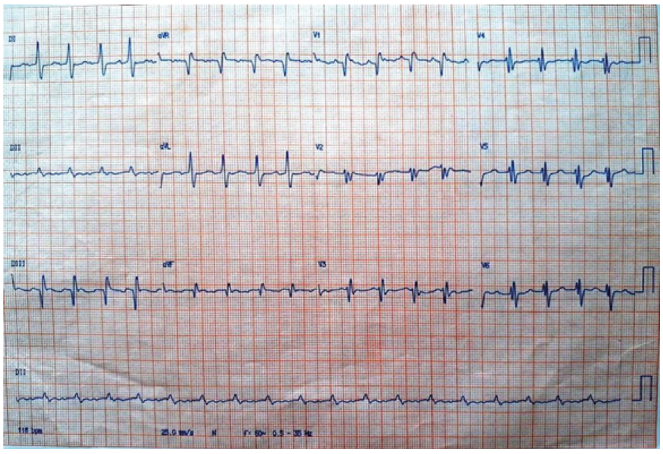


Figure 1 – *Electrocardiogram showing atrial tachycardia, complete right bundle branch block, low diffuse voltage, and signs of right ventricular overload.*

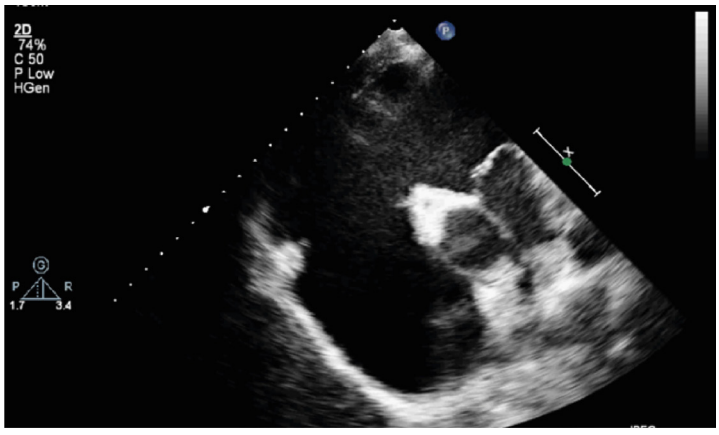


Figure 2 – *Transthoracic echocardiogram of the parasternal short-axis window showing remnants of the septal and anterior leaflets of the tricuspid valve.*

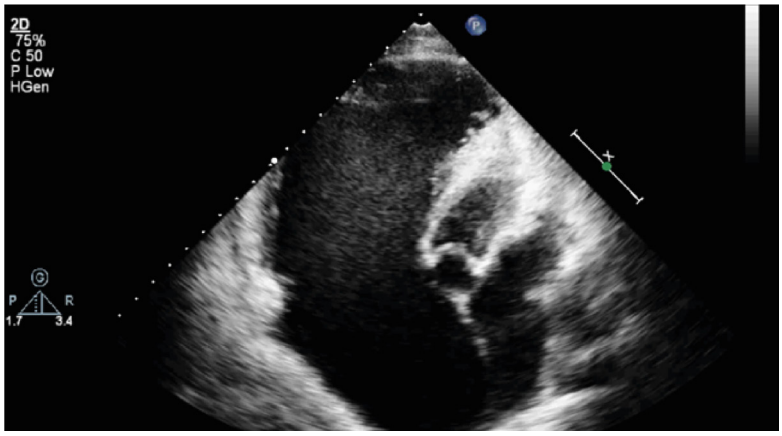


Figure 3 – *Transthoracic echocardiogram of the parasternal short-axis window showing tricuspid leaflet remnants, normal valve implantation, and right chamber dilation.*

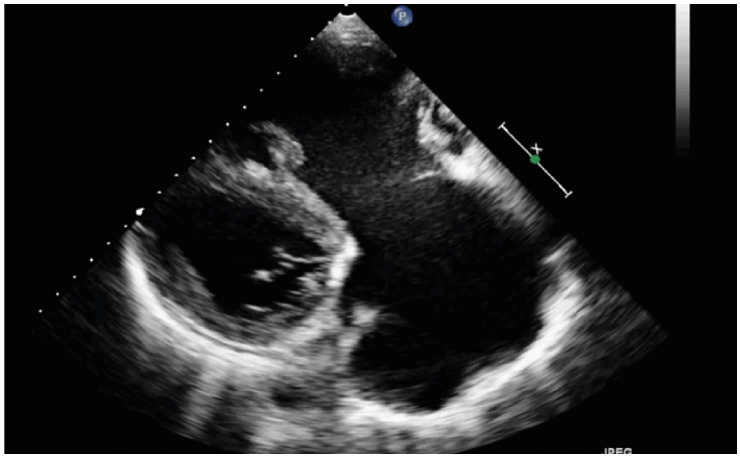
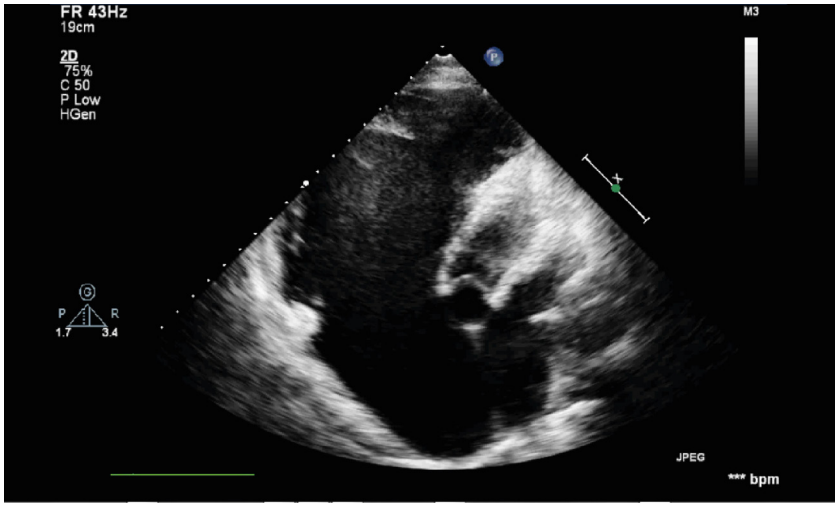
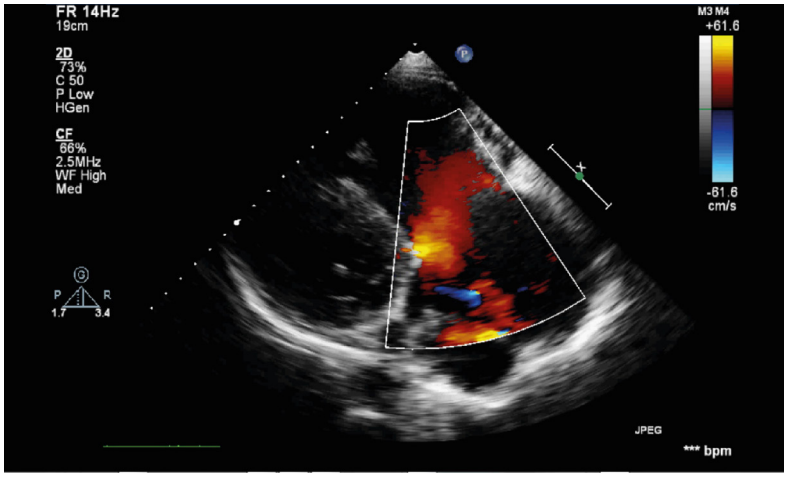


Figure 4 – *Transthoracic echocardiogram of the parasternal longitudinal window with better visualization of the right cavities showing the anterior leaflet of the tricuspid valve with a normal implant in the atrioventricular groove.*



Video 1 – *Transthoracic echocardiogram — Parasternal short-axis view showing remnants of tricuspid leaflets and normal valve implantation, in addition to right chamber dilation.*



Video 2 – *Transthoracic echocardiogram — Longitudinal parasternal view with better image of the right cavities, showing anterior leaflet of tricuspid valve with normal implantation in the atrioventricular sulcus revealing its flow on color Doppler.*

anomaly differs from UTO since it shows lesser displacement of the tricuspid septal leaflet on the echocardiography examination that can present varying degrees of adherence in the septal region and the presence of an anterior cuspid leaflet.[6]

Enlarged right heart chambers are related to the natural history of congenital UTO, being an indicator of poor prognosis when the dilation is severe as well as pulmonary hypoplasia and relative hypoplasia of the pulmonary artery trunk.[7] Many patients tolerate tricuspid regurgitation well and become symptomatic only in adulthood, which decreases the use of surgical treatment since its natural history is quite variable and surgical results have not shown to be significant.[2,3,7] The main therapeutic choice is using diuretics to treat systemic venous congestion[3] as in the present case.

Conclusion

The findings of the present case report suggest that congenital UTO, despite being a very rare entity, must be part of the diagnostic hypotheses for heart diseases developing with RHF, especially in patients with an undefined etiology, associated with typical echocardiography changes. Thus, advanced diagnostic methods, particularly echocardiography, have contributed to the early and accurate diagnosis of rare congenital cardiac malformations.

Conflict of interest

The authors have declared that they have no conflict of interest.

References

1. Kumar Vikraman S, Chandra V, Balakrishnan B, Jaiman S, Batra M, Kannoly G. Unguarded tricuspid orifice-a rare cause of fetal right atrial dilatation with characteristic color doppler sign: Case report with review of literature. J Clin Ultrasound. 2017;45(6):370-374.
2. Mohan JC, Tomar D, Shekhar C. Congenitally unguarded tricuspid valve orifice with multiple other defects in a child with refractory heart failure. Indian Heart J. 2009;61(1):89-92.
3. Abdelnabi MH, Shehata H, Saleh Y, Elgowelly M, Almaghraby A. A case of unguarded tricuspid valve orifice diagnosed accidentally in an adult. Echocardiography. 201;36(3):615-617.
4. Gupta A, Grover V, Gupta VK. Congenital tricuspid regurgitation: review and a proposed new classification. Cardiol Young. 2011;21(2):121-9.
5. Ozkutlu S, Günal N, Cağlar M, Alehan D, Güngör C. Unguarded tricuspid orifice: a rare malformation of tricuspid valve diagnosed by echocardiography. Report of two cases and review of the literature. Int J Cardiol. 1996;56(2):125-9.
6. Kanjuh VI, Stevenson JE, Amplatz K, Edwards JE. Congenitally unguarded tricuspid orifice with coexistent pulmonary atresia. Circulation. 1964;30:911-7.
7. Mohan JC, Shukla M, Mohan V, Sethi A. Congenitally unguarded tricuspid valve orifice with right ventricular apical isolation in an adult. Indian Heart J. 2016;68 Suppl 2(Suppl 2):S121-S125.
8. Mohan JC, Passey R, Arora R. Unguarded tricuspid orifice and patent right ventricular outflow tract presenting with long-standing severe right failure in an adult. International Journal of Cardiology [Internet]. 1998 [cited 2020 Jun 22];66:85-87. https://www.internationaljournalofcardiology.com/article/S0167-5273(98)00150-8/abstract
9. Mohan JC, Passey R, Arora R. Echocardiographic spectrum of congenitally unguarded tricuspid valve orifice and patent right ventricular outflow tract. International Journal of Cardiology. 2000;74:153-157.