Cardiology and Angiology: An International Journal

Volume 13, Issue 1, Page 1-4, 2024; Article no.CA.111101
ISSN: 2347-520X, NLM ID: 101658392

Isolated Double-Chambered Right Ventricle: A Rare Congenital Heart Disease

Sofia Bezza [a*], Mohamed Imad Rhoujjati [a], Mohammed Eljamili [a], Saloua El Karimi [a] and Mustapha Elhattaoui [a]

[a] Department of Cardiology, Mohammed VI University Hospital, Marrakesh, Morocco.

Authors' contributions

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

Article Information

DOI: 10.9734/CA/2024/v13i1385

Open Peer Review History:
This journal follows the Advanced Open Peer Review policy. Identity of the Reviewers, Editor(s) and additional Reviewers, peer review comments, different versions of the manuscript, comments of the editors, etc are available here: https://www.sdiarticle5.com/review-history/111101

Case Study

Received: 23/10/2023
Accepted: 28/12/2023
Published: 03/01/2024

ABSTRACT

Double-chambered right ventricle (DCRV) is an uncommon congenital heart condition characterized by a progressive obstruction in the right ventricular outflow tract. This obstruction is caused by anomalous muscles or fibrous tissues that divide the right ventricle into two cavities: a proximal high-pressure chamber (anatomically lower) and a distal low-pressure chamber (anatomically higher). We present a case of a middle-aged man with a medical history of recurrent symptomatic dyspnea. Upon presentation, there were no signs of congestive cardiac failure. The diagnosis was confirmed using transthoracic two-dimensional (2-D) echocardiography and Transesophageal echocardiography (TEE) for more precise characterization. The primary objective of this case report is to highlight the rarity of this congenital heart disease, particularly in adults.

**Corresponding author: E-mail: bezzasf.fmpm@gmail.com;*

Keywords: *Double-chambered right ventricle; adult congenital heart disease; right ventricular outflow tract obstruction; echocardiography.*

1. INTRODUCTION

Double-chambered right ventricle (DCRV) is an uncommon congenital heart disease marked by the division of the right ventricular cavity into two chambers by anomalous muscle bundles [1].

Typically, DCRV is diagnosed at childhood or adolescence, and most DCRV patients have associated congenital anomalies, such as ventricular septal defect (VSD), pulmonary stenosis, and subaortic stenosis [2]. Here, we report a case of an isolated DCRV in an adult patient

2. PRESENTATION OF CASE

We describe the case of a 53-year-old man, with no known personal history of disease or previous diagnosis of a congenital heart disease, who was admitted due to a progressive shortness of breath over the last 4 months. Upon examination, he's heart rate was 90beats/min ,the blood pressure was 107/57 mHg and the respiratory rate at 20 breaths per minute. On physical examination the extremities were well perfused with strong and symmetric pulses with no sign of left or right heart failure. the cardiac auscultation noted a systolic ejection murmur at the left parasternal border. The ECG displayed right ventricular hypertrophy with right axis deviation, indicative of right ventricular overload. A chest x-ray confirmed cardiomegaly with a 0.7 cardiothoracic ratio.

Two-dimensional (2D) transthoracic echocardiography was performed revealing a turbulent Doppler color flow jet indicative of a stenotic mid-right ventricle (Fig. 1). Continuous-wave Doppler analysis across this turbulent jet exhibited flow acceleration measuring 4.53 m/s, corresponding to a pressure gradient of approximately 82 mmHg, as calculated using the simplified Bernoulli equation. It is important to note that the obstructive gradient might have been underestimated due to the presence of severe tricuspid regurgitation. Additionally, the examination identified right atrial enlargement, right ventricular dilation, moderate dilatation of the pulmonary artery with normal infundibular contraction, and severe tricuspid regurgitation with a dual primitive and hemodynamic component exacerbated by right ventricular mid-ventricular stenosis

Transesophageal echocardiography (TEE) was performed for a better analysis and demonstrated an anomalous muscle bundle dividing the RV into two parts, with turbulent flow jet on color Doppler images

3. DISCUSSION

A double-chambered right ventricle (DCRV) is a congenital cardiac anomaly characterized by the division of the right ventricle into two chambers. Anomalous muscles or fibrous tissues within the right ventricular cavity create a proximal high-pressure (anatomically lower) chamber and a distal low-pressure (anatomically higher) chamber. The incidence of DCRV accounts for 0.5%-2% of all cases of congenital heart disease [3,2].

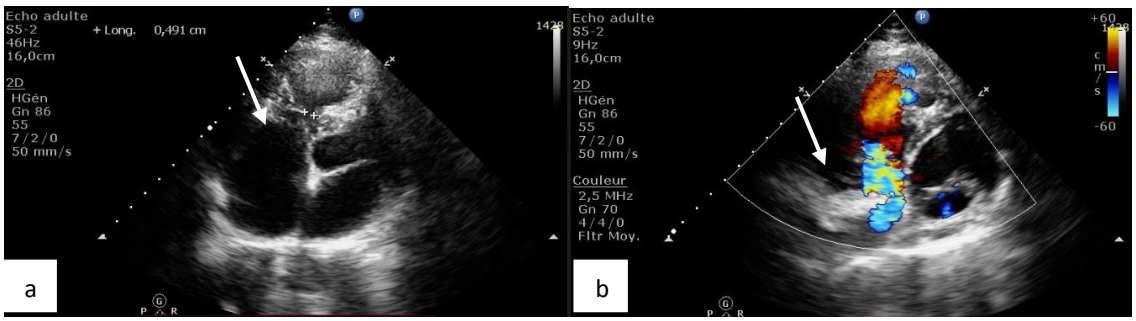

Fig. 1. Transthoracic echocardiogram on the parasternal short axis view showing stenotic mid-right ventricle with an anomalous muscle bundle as seen in systole; (b) Doppler color showing moderate to severe tricuspid regurgitation between the right atrium and right ventricle

The finding of a DCRV is exceptionally rare as an isolated anomaly. It is more commonly associated with a membranous type ventricular septal defect (VSD). Other reported associated lesions include: Subaortic stenosis, pulmonary valve stenosis, double outlet right ventricle, tetralogy of Fallot, anomalous pulmonary venous drainage, complete or corrected transposition of the great arteries, pulmonary atresia with intact ventricular septum and Ebstein's anomaly of the tricuspid valve [2,4].

The anomalous muscle bundles, varying in number from 1 to many, typically originate from the body of the septal band (septomarginal trabeculation) and traverse the right ventricular chamber to reach the anterior free wall [5,6].

A simple classification of the pathology was proposed by Folger, who described two positions of the abnormal muscle bundle: high (or horizontal) and low (or oblique).

Most cases of DCRV are diagnosed and treated during childhood, where as an initial presentation during adulthood has been reported in rare cases [7,2,8].

A similar type of case report has been published by Park et al., in which they reported a case of isolated DCRV presenting in an asymptomatic person“ [3]. One another case report was published by Pansari et al., in which they reported a case of isolated DCRV presenting with congestive heart failure [9], another case report was published by Ashok Garg,et al in which ther reported a case of isolated DCRV in a 18-year-old male [10], a rare case report of isolated DCRV with dextrocardia. was published [11].

Presenting symptoms are usually due to severe RVOT obstruction leading to a low cardiac output state or due to associated right heart failure.

Although transthoracic echocardiography (TTE) is widely accepted for demonstrating double-chambered right ventricle (DCRV) in pediatric patients, challenges may arise in studying adults. The implementation of transesophageal echocardiography (TOE) provides better lesion definition in adults. In certain cases, alternative modalities such as magnetic resonance or cardiac catheterization may be necessary [12].

The timing of surgical repair depends on the presence of associated cardiac anomalies. If there are no other significant coexisting defects,, observation is possible as long as the intracavitary pressure gradient is not more than 40 mmHg and the obstruction is not progressive [13].

Surgical correction consists of resection of the AMB and closure of the VSD through a transatrial approach to achieve adequate exposure of the AMB. Surgery offers complete relief of the obstruction, and a substantial improvement in symptoms and functional status. The long-term results of surgical treatment are excellent [14].

Postoperative complications have been documented in the literature, specifically ventricular arrhythmias mitigated through medical intervention, and instances of complete right bundle branch block. In cases where there was no progression to high-degree ventricular arrhythmia disorders, permanent pacing was deemed unnecessary,

According to Ragiab et al, no cases of ventricular tachycardia were reported in the study supraventricular tachycardia was found in only one patient, who was successfully treated with a beta blocker. Similarly, Kveselis et al reported a relatively low incidence of cardiac complication after surgical repair.

In view of these results documented in the literature, Surgical repair of a double-chambered right ventricle yields excellent haemodynamic and functional results, Nevertheless mid- and long-term surveillance is necessary [15,16,17].

4. CONCLUSION

Double-chambered right ventricle (DCRV), which leads to obstruction in the right ventricular outflow tract, is an uncommon congenital heart disease, particularly when initially diagnosed in adults. The diagnosis can be challenging, especially when it manifests with atypical symptoms. The majority of DCRV cases are associated with congenital anomalies, with ventricular septal defect (VSD) or fixed subaortic stenosis being the most common. An isolated DCRV, without associated anomalies, is exceptionally rare, Surgical repair of a double-chambered right ventricle yields excellent haemodynamic and functional results over the mid to long term.

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.

REFERENCES

1. Garg A, Agrawal D, Sharma GL. Isolated double-chambered right ventricle–A rare entity. Journal of Cardiovascular Echography. 2020;30(3):162.
2. Park JI, Kim YH, Lee K, Park HK, Park CB. Isolated double-chambered right ventricle presenting in adulthood. Int J Cardiol. 2007;121(3):25‑7.
3. Park JG, Ryu HJ, Jung YS, Kim KJ, Lee BR, Jung BC, et al. Isolated Double-Chambered Right Ventricle in a Young Adult. Korean Circ J. Mai 2011;41(5): e272‑5.
4. Folger GM. Right ventricular outflow pouch associated with double-chambered right ventricle. Am Heart J. Mai 1985;109(5): 1044‑9.
5. Mohsen A, Rahman F, Ikram S. Anomalous muscle bundles causing double-chambered right ventricle in adults. J Invasive Cardiol. 2013;25(12):E212-213.
6. El Kouache M, El Houari T, Fellat N, Benomar M. Right mid-ventricular stenosis with intact interventricular septum. Int J Cardiol. 2007;122(2):e13‑4.
7. Lee WJ, Song BG, Kang GH, Park YH, Chun WJ, Oh JH. A case of asymptomatic isolated double-chambered right ventricle in an adult man. J Clin Ultrasound JCU. 2013;41(9):579‑81.
8. Mohamed S, Amine K, Souad A, Rachida A, Mohamed C. Double chambered right ventricle with an intact interventricular septum: Case report.
9. Pansari N, Raghavendra H, Mahur H, Dave M. Double Chambered Right Ventricle: A Rare Diagnosis. J Assoc Physicians India. Mars. 2017;65(3):96‑8.
10. Garg A, Agrawal D, Sharma GL. Isolated Double-Chambered Right Ventricle – A Rare Entity. J Cardiovasc Echography. 2020;30(3):162‑4.
11. Double-Chambered Right Ventricle and Situs Inversus With Dextrocardia | Circulation [Internet]. Disponible sur. Available:https://www.ahajournals.org/doi/full/10.1161/CIR.0b013e3181d56ebd [Access on 22 Oct 2023].
12. Hoffman P, Wójcik AW, Rózański J, Siudalska H, Jakubowska E, Włodarska EK, et al. The role of echocardiography in diagnosing double chambered right ventricle in adults. Heart Br Card Soc. juill 2004;90(7):789‑93.
13. McElhinney DB, Chatterjee KM, Reddy VM. Double-chambered right ventricle presenting in adulthood. Ann Thorac Surg.. 2000;70(1):124‑7.
14. Babakhoya A, Labib S, El Madi A, Atmani S, Harandou M, Abouabdilah Y, et al. Repair of isolated double-chambered right ventricle. Afr J Paediatr Surg. 2013;10(2): 199.
15. Telagh R, Alexi-Meskishvili V, Hetzer R, Lange PE, Berger F, Abdul-Khaliq H. Initial clinical manifestations and mid- and long-term results after surgical repair of double-chambered right ventricle in children and adults. Cardiol Young. 2008;18(3): 268‑74.
16. Kottayil BP, Dharan BS, Pillai VV, Panicker VT, Gopalakrishnan SK, Jayakumar K. Surgical repair of double-chambered right ventricle in adulthood. Asian Cardiovasc Thorac Ann. févr 2011;19(1): 57‑60.
17. Kveselis D, Rosenthal A, Ferguson P, Behrendt D, Sloan H. Long-term prognosis after repair of double-chamber right ventricle with ventricular septal defect. Am J Cardiol. 1984;54(10):1292‑5.

Peer-review history:
The peer review history for this paper can be accessed here:
https://www.sdiarticle5.com/review-history/111101