Isolated Double-chambered Right Ventricle:a rare congenital heart disease

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.

*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”. [17] “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” .(1) 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 (Figure 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 regurgitationwith 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

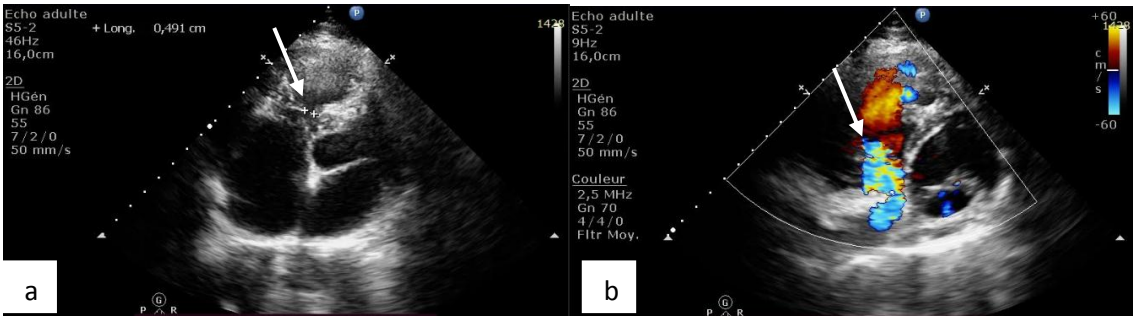

Figure 1:Transthoracicechocardiogram on the parasternal short axis viewshowing stenoticmid-right ventriclewith an anomalous muscle bundle as seen in systole ;(b) Doppler colorshowingmoderate to severetricuspidregurgitationbetween the right atrium and right ventricle

3. DISCUSSION :

“A double-chambered right ventricle (DCRV) is a congenitalcardiacanomalycharacterized by the division of the right ventricleintotwochambers. Anomalous muscles or fibrous tissues within the right ventricularcavitycreate a proximal high-pressure (anatomicallylower) chamber and a distal low-pressure (anatomicallyhigher) chamber. The incidence of DCRV accounts for 0.5%-2% of all cases of congenitalheartdisease“. (2)(1)
“The finding of a DCRV isexceptionally rare as an isolatedanomaly. It is more commonlyassociatedwith a membranous type ventricular septal defect (VSD). Otherreportedassociatedlesionsinclude: subaorticstenosis, pulmonary valve stenosis, double outlet right ventricle, tetralogy of Fallot, anomalouspulmonaryvenous drainage, complete or corrected transposition of the greatarteries, pulmonaryatresiawith intact ventricular septum and Ebstein'sanomaly of the tricuspid valve“(1)(3)

"The anomalous muscle bundles, varying in numberfrom 1 to many, typicallyoriginatefrom the body of the septal band (septomarginaltrabeculation) and traverse the right ventricularchamber to reach the anterior free wall".(4)(5)
"A simple classification of the pathologywasproposed by Folger, whodescribedtwo positions of the abnormal muscle bundle: high (or horizontal) and low (or oblique)
Most cases of DCRV are diagnosed and treatedduringchildhood, whereas an initial presentationduringadulthood has been reported in rare cases"(6)(1)(7)
"A similar type of case report has been published by Park et al., in whichtheyreported a case of isolated DCRV presenting in an asymptomaticperson" , (2) One another case report waspublished by Pansari et al., in whichtheyreported a case of isolated DCRV presentingwith congestive heartfailure.(8), another case report waspublished by Ashok Garg,et al in whichtherreported a case of isolated DCRV in a 18-year-old male(9), a rare case report of isolated DCRV withdextrocardia. waspublished(10)
Presentingsymptoms are usually due to severe RVOT obstruction leading to a lowcardiac output state or due to associated right heartfailure.
" Althoughtransthoracicechocardiography (TTE) iswidelyaccepted for demonstrating double-chambered right ventricle (DCRV) in pediatric patients, challenges may arise in studyingadults. The implementation of transesophagealechocardiography (TOE) providesbetterlesiondefinition in adults. In certain cases, alternative modalitiessuch as magneticresonance or cardiaccatheterizationmaybenecessary" . (11)
"The timing of surgicalrepairdepends on the presence of associatedcardiac anomalies. If there are no othersignificantcoexistingdefects,, observation is possible as long as the intracavitary pressure gradient is not more than 40 mmHg and the obstruction is not progressive" .(12)
"Surgical correction consists of resection of the AMB and closure of the VSD through a transatrialapproach to achieveadequateexposure of the AMB. Surgeryofferscomplete relief of the obstruction, and a substantialimprovement in symptoms and functionalstatus. The long-termresults of surgicaltreatment are excellent"(13)
Postoperative complications have been documented in the literature, specificallyventriculararrhythmiasmitigatedthroughmedical intervention, and instances of complete right bundle branch block. In cases wheretherewas no progression to high-degreeventriculararrhythmiadisorders, permanent pacingwasdeemedunnecessary,
According to Ragiab et al, no cases of ventriculartachycardiawerereported in the studysupraventriculartachycardiawasfound in only one patient, whowassuccessfullytreatedwith a beta blocker. Similarly, Kveselis et al reported a relativelylow incidence of cardiac complication aftersurgicalrepair.

"In view of theseresultsdocumented in the literature, Surgicalrepair of a double-chambered right ventricleyields excellent haemodynamic and functionalresults, Neverthelessmid- and long-term surveillance isnecessary".(14)(15)(16)

4. CONCLUSION :

Double-chambered right ventricle (DCRV), which leads to obstruction in the right ventricularoutflow tract, is an uncommoncongenitalheartdisease, particularlywheninitiallydiagnosed in adults. The diagnosis can bechallenging, especiallywhenitmanifestswithatypicalsymptoms. The majority of DCRV cases are associatedwithcongenital anomalies, withventricular septal defect (VSD) or fixedsubaorticstenosisbeing the mostcommon. An isolated DCRV, withoutassociated anomalies, isexceptionally rare, Surgicalrepair of a double-chambered right ventricleyields excellent haemodynamic and functionalresults 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 writtenethicalapproval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declaredthat no competinginterestsexist

REFERENCES

1. Park JI, Kim YH, Lee K, Park HK, Park CB. Isolated double-chambered right ventricle presenting in adulthood. Int J Cardiol. 18 oct 2007;121(3):e25‑7.

2. 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):272‑5.

3. Folger GM. Right ventricular outflow pouch associated with double-chambered right ventricle. Am Heart J. mai 1985;109(5):1044‑9.

4. Mohsen A, Rahman F, Ikram S. Anomalous muscle bundles causing double-chambered right ventricle in adults. J Invasive Cardiol. déc 2013;25(12):E212-213.

5. El Kouache M, El Houari T, Fellat N, Benomar M. Right mid-ventricular stenosis with intact interventricular septum. Int J Cardiol. nov 2007;122(2):e13‑4.

6. 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.

7. Mohamed S, Amine K, Souad A, Rachida A, Mohamed C. Double chambered right ventricle with an intact interventricular septum: Case report.

8. 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.

9. Garg A, Agrawal D, Sharma GL. Isolated Double-Chambered Right Ventricle – A Rare Entity. J Cardiovasc Echography. 2020;30(3):162‑4.

10. Double-Chambered Right Ventricle and Situs Inversus With Dextrocardia | Circulation [Internet]. [cité 22 oct 2023]. Disponible sur: https://www.ahajournals.org/doi/full/10.1161/CIR.0b013e3181d56ebd

11. 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.

12. McElhinney DB, Chatterjee KM, Reddy VM. Double-chambered right ventricle presenting in adulthood. Ann Thorac Surg. juill 2000;70(1):124‑7.

13. 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.

14. 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. juin 2008;18(3):268‑74.

15. 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.

16. 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. 1 déc 1984;54(10):1292‑5.

17. Garg A, Agrawal D, Sharma GL. Isolated double-chambered right ventricle–a rare entity. Journal of Cardiovascular Echography. 2020 Jul;30(3):162.