# Un cas de ventricule droit à double chambre chez un adulte.

# A case of isolated double-chambered right ventricle in an adult

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## Résumé

Le ventricule droit à double chambre (VDDC) est une malformation cardiaque rare aboutissant à une obstruction médio-ventriculaire droit. Son diagnostic est généralement fortuit chez les patients adultes. Nous présentons ici le cas d'une femme de 60 ans sans antécédents cardiaques qui a présenté des palpitations à l'effort. L'électrocardiogramme a montré des extrasystoles ventriculaires naissant du ventricule droit. L'échocardiographie transthoracique a révélé une obstruction médio-ventriculaire droite par un muscle anormal hypertrophié divisant le ventricule droit en deux chambres, inlet et outlet. Le diagnostic de VDDC a été confirmé par l'imagerie par résonance magnétique cardiaque. L'ablation chirurgicale du muscle hypertrophié a été initialement proposée mais refusée par la patiente. La patiente a été mise sous vérapamil pour des extrasystoles ventriculaires.

Mots-clés

Extrasystoles ventriculaires, ventricule droit, obstruction médio-ventriculaire, muscle hypertrophié

## Summary

Double-chambered right ventricle (DCRV) is a rare heart anomaly leading to right midventricular obstruction. It is usually an incidental diagnosis in adult patients. Herein we present the case of a 60-year-old woman with no cardiac history who presented with palpitations on exertion. Electrocardiogram showed ventricular extrasystoles originating in the right ventricle. Transthoracic echocardiography revealed right midventricular obstruction by a hypertrophic abnormal muscle dividing the right ventricle into two chambers, inlet and outlet. The diagnosis of DCRV was confirmed by cardiac magnetic resonance imaging. Surgical excision of the hypertrophic muscle was initially proposed but refused by the patient. The patient was put on verapamil for ventricular extrasystoles.

Keywords

Ventricular extrasystoles, right ventricule, midventricularobstruction, hypertrophic muscle

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## INTRODUCTION

Double-chambered right ventricle (DCRV) is a rare cardiac anomaly [1]. It is characterized by a right ventricle (RV) divided by an aberrant hypertrophic muscle into two chambers, proximal or inlet and distal or outlet [2]. DCRV is often associated with other cardiac malformations such as ventricular septal defect (VSD) [1]. The diagnosis is usually made in infancy, rarely in adulthood. Depending on the extent of right midventricular obstruction, patient can be asymptomatic or complaining of non-specific symptoms. Herein we present a case of DCRV diagnosed by transthoracic echocardiography (TTE) and magnetic resonance imaging (MRI).

## CASE PRESENTATION

A 60-year-old woman was received in cardiology clinics for complaints of palpitations on exertion evolving over 2 years. She had no medical history and she was normotensive. There was an intense heart murmur in the left sternal border with no signs of left or right heart failure. Transcutaneous oxygen saturation was 98%. Electrocardiogram showed sinus rhythm with incomplete right bundle branch block associated to ventricular extrasystoles with left bundle branch block morphology and inferior axis. TTE showed bilateral and located hypertrophic muscle bands next to the pulmonary infundibulum, arising from both inner faces of the ventricular septum and the RV anterior wall and extending over 15 mm in length. These muscle bands were responsible for RV outflow tract narrowing to a diameter of 6 mm and thus dividing the RV into two chambers (figure 1). There was no attachment of muscle bands on the tricuspid pillars. Color Doppler flow showed a turbulent and mosaic flow in the narrowed RV outflow suggestive of significant RV outflow diameter reduction. Continuous Doppler flow demonstrated a proximal high-pressure chamber (121 mmHg) under the tricuspid valve and a distal low-pressure chamber in the distal chamber next the pulmonary infundibulum (figure 2). RV systolic function was normal. Right atrium area was estimated at 25 $cm^2$ with a mild tricuspid regurgitation. Neither VSD nor pulmonary stenosis was detected. MRI was performed to confirm the diagnosis and to rule out RV hypertrophic cardiomyopathy (figure 3). Twenty-four-hour holter monitoring showed 7% of left bundle branch block morphology ventricular extrasystoles suggestive of the right ventricle origin with no malignant electrical signs. Surgical excision of the hypertrophic muscle was initially proposed but refused by the patient. She was put on 240 milligrams of verapamil daily. On clinical follow-up at 3 years, palpitations have disappeared completely. Follow-up TTE showed no modification in Doppler parameters. Control twenty four hour holter showed no ventricular extrasystoles.

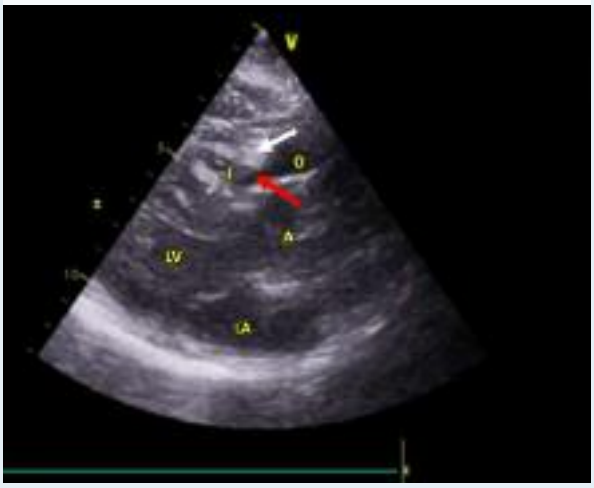


**Figure 1.** Transthoracic echocardiogram, parasternal long axis view shows right midventricular obstruction (red arrow) by hypertrophic muscle bands (white arrow). Right ventricle is divided into Inlet (I) and Outlet (O). A: Aorta, LA: left atrium, LV: left ventricle.

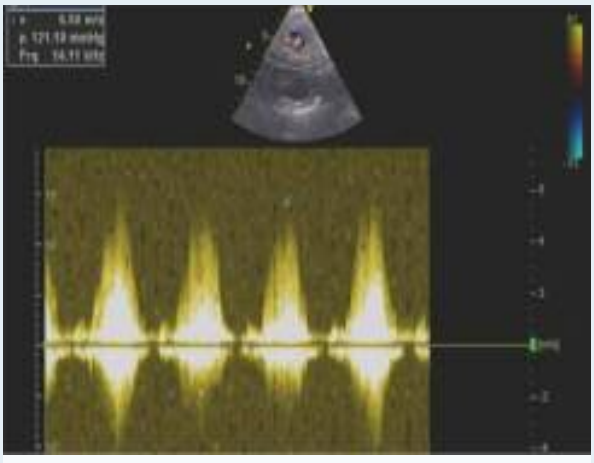

**Figure 2.** Transthoracic echocardiogram, parasternal long axis view shows Doppler peak gradientestimated at 121 mmHg in the proximal chamberunder the tricuspid valve.

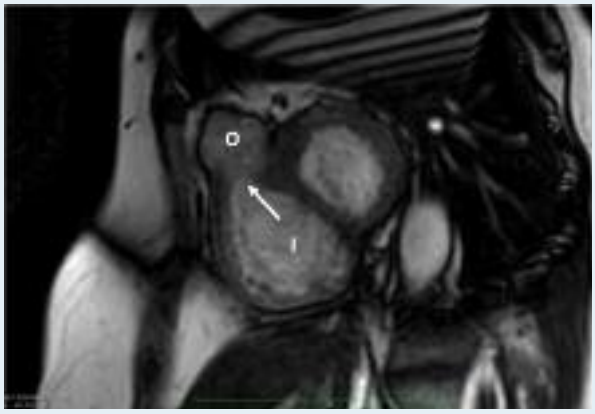

**Figure 3.**Saggital T2-weighted cardiac magnetic resonance image shows located right midventricular obstruction (white arrow) dividingthe right ventricle into two chambers, inlet (I) and outlet (O).

## DISCUSSION

DCRV is a rare cardiac anomaly that represents 0.5 to 2 % of all congenital heart diseases [1]. It was first described by Peacock in 1858 as a RV divided by an aberrant hypertrophic muscle into two chambers, proximal or inlet with high pressure and distal or outlet, with low pressure [2, 3]. Right midventricular obstruction remains poorly explained due to the reduced number of reported cases. Various mechanisms may explain the abnormal muscle hypertrophy: a hypertrophy of a muscular shelf from the septomarginal or the septoparietal trabeculations and its extension to RV apex, a displacement of moderator band and a disturbance of blood flow in RV outflow tract in patients with VSD [4, 5]. Restivo et *al* proposed subtypes of DCRV including anomalous apical shelf with or without Ebstein malformation, hypertrophy of apical trabeculations, anomalous septoparietal band and right midventricular obstruction by a circumferential muscular diaphragm in tetralogy of Fallot [6]. In 80 to 90% of cases, DCRV is associated with other heart malformations such as VSD, tetralogy of Fallot, sub-aortic stenosis, partial anomalous pulmonary venous return hence the importance of screening for such anomalies [7]. Isolated DCRV exceptionally manifests in adulthood. Patient can be asymptomatic or present with non-specific symptoms including asthenia, exertion dyspnea, cyanosis, palpitations, syncope or chest pain [8]. Clinical examination is usually is usually not very contributory. In the present case, palpitations and systolic murmur were the elements that led to diagnosis suspicion. LBBB ventricular extrasystoles morphology and inferior axis are suggestive of RV outflow tract origin. TTE is the imaging modality of choice for the diagnosis in children although less reliable in adult patients. It allows the diagnosis of DCRV in only 15% of cases because of retrosternal location and poor echogenicity of the RV [9].

Transesophageal echocardiography (TEE) is more suitable than TTE to determine the full details of the pathology and to estimate the pressure gradient [9].TEE could have limits in increased trabeculations of the RV [10]. In our case, the patient did not tolerate TEE. Right midventricular obstruction could be diagnosed by MRI showing obstructive flow and ruling out isolated RV hypertrophic cardiomyopathy [11]. Given the rarity of the diagnosis, no clear recommendations exist to dictate the best therapeutic strategy [12]. The recommended treatment is surgical ablation of the hypertrophic muscle when the stenosis is severe (Doppler peak gradient >64 mmHg) regardless of symptoms or when DCRV is associated to heart anomalies [13, 14]. Both transatrial and transventricular approaches in the surgical repair of DCRV were described. Calcium channel blockers or Beta-blockers may be beneficial to improve symptoms and exercise capacity in case of dynamic right midventricular obstruction [15].

## CONCLUSION

DCRV is very rare in adult patients. TTE and MRI are the methods of choice for diagnosis. Patients can be managed conservatively. Yet, routine follow-up with TTE and twenty-four-hour holter is warranted to detect right midventricular obstruction progression and/ or ventricular arrhythmia.

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