

A case report: multiple right ventricular diverticula with constrictive pericarditis and right heart failure

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Background

Right ventricular diverticula (RVD) are very rare congenital anomalies and their association with constrictive pericarditis is even rarer. So far, only one case has been published in literature.

Case summary

We report a case of multiple congenital RVD with constrictive pericarditis and right heart failure which was incidentally identified on surveillance computed tomography (CT) for abdominal lymphangioma. Interval CT, echocardiography, and cardiac magnetic resonance imaging (CMR) studies were performed and reviewed. Computed tomography abdomen showed hepatic congestion with features of portal hypertension, increasing size of the RVD on review of serial CTs, and eccentric foci of pericardial calcification. Echocardiography performed for breathlessness demonstrated supranormal early diastolic tissue velocities with average of 19.8 cm s^{-1} and a septal bounce phenomenon on m-mode imaging suggesting constrictive physiology, which triggered a CMR referral. Cardiac magnetic resonance imaging HASTE and right ventricular (RV) outflow tract imaging showed four outpouchings along RV free wall, the largest measuring $4.5 \times 2 \text{ cm}$ with a sizeable neck. These outpouchings displayed a trabecular network and/or were contractile aiding the diagnosis of diverticula as opposed to aneurysms. Right ventricular function was moderately compromised, whereas left ventricular function was preserved.

Discussion

Right ventricular diverticula can be associated with, and potentially be causative of, pericardial thickening and calcification eventually leading to constrictive pericarditis and heart failure.

Keywords

Right ventricular diverticulum • Ventricular aneurysm • Pericardial calcification • Constrictive pericarditis • Heart failure • Case report

Learning points

- Ventricular diverticula are rare ($\sim 0.2\%$) and more common in the left ventricle. At times ($\sim 30\%$) they are isolated incidental findings, but can be associated with congenital cardiac abnormalities, such as ventricular septal defect, tetralogy of Fallot, or tricuspid atresia.
- Ventricular diverticulum may be associated with pentalogy of Cantrell which is a syndrome comprising of congenital anomalies of the thoracic and abdominal midline, diaphragmatic and sternal defects, and partial absence of the inferoapical pericardium and it should be considered while investigating such cases.

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Introduction

Congenital ventricular diverticulum is a rare cardiac anomaly with a reported incidence of 0.013–0.016%.^{1,2} It can be found anywhere in the right or left ventricular wall; commonly in the apex and perivalvular region. Right ventricular diverticulum (RVD), however, is an extremely rare condition with fewer than 20 cases reported in the literature.^{3,4} Right ventricular diverticula can occur as an isolated anomaly or be associated with other congenital cardiac anomalies or be a part of pentalogy of Cantrell which is associated with midline abdominal wall or sternal defects, deficiency of anterior diaphragm or diaphragmatic pericardium, and congenital cardiac anomalies.⁵ We report a rare case of multiple RVD associated with constrictive pericarditis and heart failure. Our report highlights the diagnostic imaging features of RVD along with its associated anomalies and a brief literature review.

Timeline

	Presentation	Investigations	Findings
8 years prior to current admission	Fever of unknown origin	Computed tomography (CT) thorax, abdomen, and pelvis	Abdominal lymphangioma and right ventricular outpouching
1 year prior to current admission	SOB, leg swelling, and inability to lose weight	CT thorax, abdomen, and pelvis	Right ventricular diverticulum, pericardial thickening and calcification, and hepatic congestion
Current admission	Breathlessness and ascites	Echocardiography and cardiac magnetic resonance imaging	Constrictive pericarditis and features of right heart failure

Case presentation

A 53-year-old male Caucasian patient was incidentally diagnosed with multiple right ventricular outpouchings (RVOs) 8 years ago, when he was investigated for fever of unknown origin. Computed tomography (CT) scan of his chest and abdomen showed abdominal lymphangioma (Figure 1A), ascites (Figure 1B), and a RVO (Figure 1C) measuring 3 × 2.5 cm. His abdominal lymphangioma was resected surgically and he did not undergo chemo or radiotherapy. Right ventricular outpouching was left alone without any intervention for observation and monitoring over time. An anterior abdominal wall hernia (Figure 1D) was also noticed, probably a forme fruste of Cantrell syndrome. The patient developed shortness of breath, leg swelling and was unable to lose weight 7 years post-surgery. Physical examination was equivocal, and he had a repeat CT scan for chest and abdomen which revealed multiple RVOs and eccentric foci of pericardial calcification near the neck of the largest RVD but also over the left ventricle AV groove and basal lateral wall (Figure 1E). Furthermore, features of fluid overload such as diffuse oedema of abdominal wall (Figure 1F), *misty mesentery*, mild to moderate ascites, hepatic congestion, nodularity, and prominent splenic vessels were also noted.

On his latest admission with shortness of breath, abdominal distension and ascites, echocardiography demonstrated supranormal, early

diastolic tissue velocities with average of 19.8 cms⁻¹, and a septal bounce phenomenon on m-mode imaging (Figure 2) suggesting constrictive physiology which triggered a cardiac magnetic resonance imaging (CMR) referral.

Cardiac magnetic resonance imaging viability scan with gadolinium contrast demonstrated multiple (at least four) diverticula along right ventricular (RV) free wall (Figure 3A)—the largest measuring 4.5 × 2 cm with a sizeable neck (Figure 3B) and at 1.5 cm from the tricuspid valve annulus. Another smaller diverticulum (9 mm) was seen just apically from the first and the third further close to the true RV apex. In the right ventricular outflow tract view (Figure 3C), the larger diverticulum was located to the acute RV angle and the fourth diverticulum was found in the free RV wall just beneath the pulmonary valve. All these outpouchings had displayed either a trabecular network (Figure 3D) and/or a contractile behaviour that favoured the diagnosis of diverticula as opposed to aneurysms (Supplementary material online, Videos S1–S3). First pass perfusion depicted a direct contrast communication into the diverticula without evidence of extravasation into the pericardial space (Figure 3E and Supplementary material

online, Video S4). Late gadolinium enhancement revealed localized epicardial type of hyperenhancement over the left ventricular (LV) lateral wall segments (Figure 3F) corresponding to areas of decreased signal intensity on T1 mapping mimicking pericardial pocket formation (Figure 3G and H). RV function was moderately compromised, whereas LV function was preserved (Table 1).

A diagnosis of right heart failure with constrictive physiology was made based on the presence of RV diverticula, progressive pericardial thickening and calcification, and moderately compromised RV function. The patient was medically managed with loop diuretics and spironolactone—later replaced with eplerenone due to side effects. He is scheduled for periodic surveillance.

Discussion

Cardiac diverticulum is a condition, which was not known until early 18th century when O'Bryan first described it in his work; 'The anomalies of the human heart'. Its reported incidence, in association with Cantrell's pentalogy, is 5.5 per million live births. Recent limited autopsy series have implied left ventricular diverticulum to have an incidence of 0.4%,⁶ whereas a CT series have documented a frequency of 2.2%.⁷ Ventricular diverticulum can

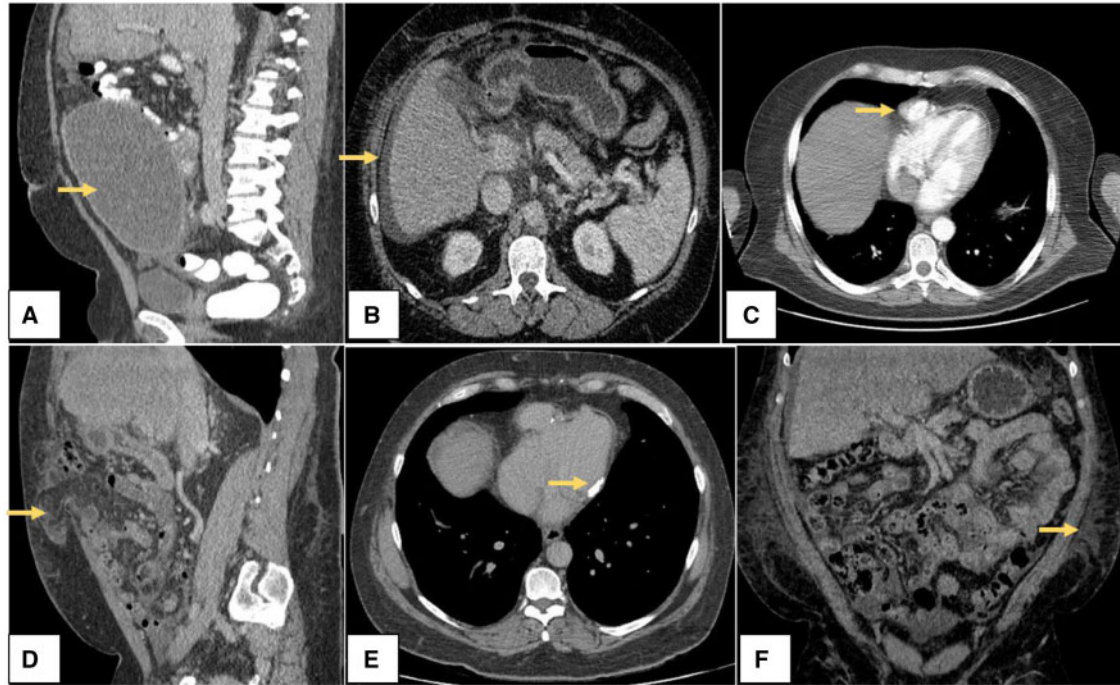


Figure 1 First, computed tomography scan 8 years ago: Arrows depict (A) abdominal lymphangioma; (B) ascites; (C) RV diverticulum; (D) anterior abdominal wall defect—second computed tomography scan 1 year ago; (E) diverticula and pericardial calcification; and (F) abdominal wall oedema.

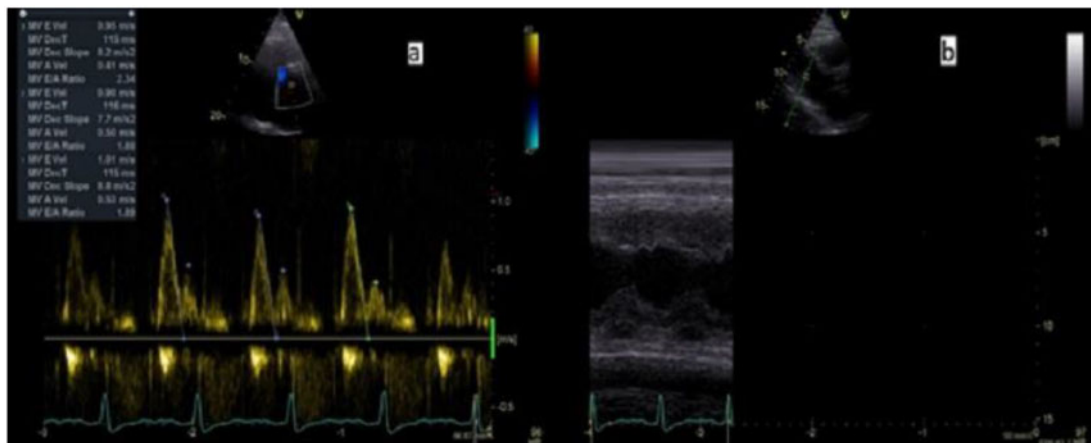


Figure 2 Echocardiography: (A) supranormal tissue velocities; and (B) septal bounce phenomenon.

be differentiated from ventricular aneurysm due to presence of a muscle layer, usually a narrow neck and synchronous contractility with the rest of the ventricle.⁸ A ventricular aneurysm being composed of only two layers, has a wide neck and may exhibit variable contractility ranging from akinesia to dyskinesia i.e. paradoxical expansion during systole and dyssynchronous contraction (reflecting elastic recoil of neighbouring myocardium) (Table 2). Aneurysms are often located apically and are associated

with thromboembolism. Congenital diverticula are considered to ensue following any antenatal ischaemic or infective insult; leading to weakening of the myocardium. However, increased ventricular pressures in constrictive pericarditis can also trigger the formation of a diverticulum and/or its subsequent expansion in later adult life. Contrarily, aneurysms are caused or associated by acute myocardial infarction, arrhythmogenic RV dysplasia, acute myocarditis, iatrogenic injury, and trauma.

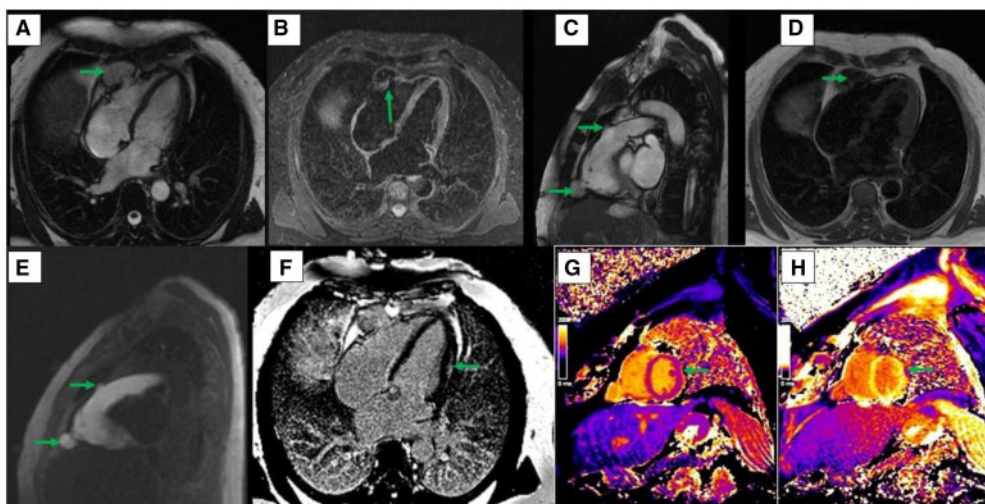


Figure 3 Gadolinium contrast enhanced cardiac magnetic resonance imaging: Arrows depict (A) still frame from cine sequence—multiple diverticula, (B) STIR BB image—neck of the diverticulum (C) right ventricular outflow tract view larger diverticulum at acute right ventricular angle and smaller diverticulum just beneath the pulmonary valve; (D) T1 BB image—trabecular network; (E) first pass perfusion—simultaneous filling of the diverticulum; (F) late gadolinium enhancement revealed localized pericardial and epicardial type of hyperenhancement over the left ventricular lateral wall segments; (G) T1 maps pre-contrast; (H) T1 maps post-contrast (decreased T1 values).

Table 1 Volumetric analysis corrected for body surface area of 2.1 m²

CMR	Right ventricle	Left ventricle
Ejection fraction (EF)	34% (49–73%)	62% (59–83%)
End diastolic volume (EDV)	174 mL or 83 mL/m ² (67–111 mL/m ²)	95 mL or 47 mL/m ² (53–97 mL/m ²)
End systolic volume (ESV)	114 mL or 54 mL/m ² (20–48 mL/m ²)	36 mL or 17 mL/m ² (10–34 mL/m ²)
Stroke volume (SV)	60 mL or 28 mL/m ² (39–71 mL/m ²)	59 mL or 28 mL/m ² (37–69 mL/m ²)

Table 2 Differences between cardiac diverticula and aneurysms

	Diverticulum	Aneurysm
Size	Small	Large
Wall	Muscular	Fibrous
Neck	Variable, narrow	Wide
Appearance	Trabeculated	Smooth
Calcification	Pericardial calcification	Thinned aneurysmal wall calcification
Contractility	Synchronous	Paradoxical, dyskinesia, and akinesia
Location	Apical and subvalvular	Apical and anterolateral
Cause	Mostly congenital	Mostly acquired
Complications	Pericarditis and cardiac failure	Thrombus, thromboembolism, and cardiac failure

Cardiac outpouchings (encompassing diverticula and aneurysms) are associated with conotruncal anomalies, annulus fibrosus, isolated ventricular septal defect, pericardial effusion, and midline thoracic and abdominal defects of the pentology of Cantrell. In 30% of cases, ventricular diverticulum can be an isolated finding.⁹ So far, only one

case has been published regarding RV diverticulum associated with constrictive pericarditis¹⁰ and this is the second case; in which, the patient developed RV dysfunction and constrictive physiology over the time along with hepatic manifestations and portosystemic shunt. However, his left ventricular function was preserved.

Patients with ventricular diverticula can remain asymptomatic throughout their lives. Conversely, they may present as pericardial effusion which usually resolves spontaneously in the absence of any haemodynamic impairment, arrhythmias, congestive cardiac failure, and rupture of the diverticulum with haemopericardium and haemodynamic compromise. In foetal life, large diverticula associated with pericardial effusion may lead to pulmonary hypoplasia.⁴

Right ventricular diverticula can be an incidental finding which can also be diagnosed on antenatal scanning; until recently six cases have been reported on antenatal scanning.¹¹ The accuracy of CMR has been reported up to 93% in differentiating constrictive from restrictive cardiomyopathy.¹⁰ On echocardiography, supranormal early diastolic tissue velocities >8 cm/s are 95% sensitive and 96% specific of constriction.¹² Calcification associated with constrictive pericarditis is better depicted with CT scan.

Management of patients with RVD varies to great extent and depends upon clinical presentation. Their resection and myocardial augmentation with pericardial fat is advocated to avert the risk of rupture and haemopericardium if these are diagnosed in early life. However, incidental diagnosis in adult life mandates long-term follow-up.¹³

Incidence of RV diverticulum is anticipated to rise due to increased availability of CMR for routine imaging both at national and international levels. Although, some work has been conducted in recent years to evaluate its incidence on cross-sectional imaging and post-mortem studies, further research is required to assess its true incidence and its impact on patients with compromised cardiac function.

Conclusion

Ventricular diverticula can be diagnosed incidentally on cross-sectional imaging. However, their precise characterization and depiction of associations/complications warrant functional imaging.

Lead author biography



Dr Sohail Iqbal, the author of many case reports served as a Specialist Radiologist in Ministry of Health Saudi Arabia for 5 years and worked in various NHS organizations as a Speciality Doctor. Currently, he has been working as a Cardiac Imaging Fellow in North West Heart Center, Manchester.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

References

1. Katsura D, Hayashi K, Tsuji S, Ono T, Ishiko A, Takahashi K, Murakami T. Fetal right ventricular diverticulum detected by prenatal ultrasound screening. *Case Rep Obstet Gynecol* 2016;6382920.
2. Sun Y, Li RJ, Xue J, Yang Y. Congenital left ventricular diverticulum diagnosed by echocardiography. *Chin Med J* 2013;126:1137.
3. Di Sessa TG, Howard SC, Salim MA. Congenital right ventricular diverticulum associated with a ventricular septal defect: a rare echocardiographic finding. *Echocardiography* 2006;23:787.
4. Caldaroni F, Rose D, Caiaro A, Casciani E, Martucci V, Manganaro L, Ventriglia F, Miraldi F. Fetal right ventricular diverticulum with pericardial effusion: to drain or not to drain? *J Pediatr Neonatal Care* 2014;1: doi:10.15406/jpnc.2014.01.00016.
5. Bellotti CA, Stewart RD, Golden AB, Mavroudis C, Vachhani N. Pentalogy of Cantrell: surgical resection of a biventricular diverticulum. *World J Pediatr Congenit Heart Surg* 2012;3:402–405.
6. Deng Y, Sun Z, Dong N, Du X. Congenital cardiac diverticulum in the subaortic valve area. *J Thorac Cardiovasc Surg* 2006;132:1087–1091.
7. Srichai MB, Hecht EM, Kim DC, Jacobs JE. Ventricular diverticula on cardiac CT: more common than previously thought. *Am J Roentgenol* 2007;189: 204–208.
8. Capasso R, Panelo M, Fiorelli A, Carbone I, Galea N. Diverticulum, or not diverticulum, that is the question! Discussing about a case of left ventricular out-pouching associated with bicuspid aortic valve assessed by cardiac magnetic resonance. *J Cardiovasc Thorac Res* 2015;7:72–74.
9. Gruberg L, Goldstein SA, Pfister AJ, Monsein LH, Evans DM, Leon MB. Images in cardiovascular medicine. Cantrell's syndrome: left ventricular diverticulum in an adult patient. *Circulation* 2000;101:109–110.
10. Ocak I, Turkbey B, Lacomis JM. Constrictive pericarditis presenting with an out-pouching of the right ventricle free wall simulating an aneurysmal dilatation. *Br J Radiol* 2011;84:e058–e061.
11. Wax JR, Moran A, Pinette MG, Reyes A, Cartin A, Blackstone J. Prenatal sonographic diagnosis of fetal right ventricular diverticulum. *J Ultrasound Med* 2007;26: 267–270.
12. Ha J-W, Ommen SR, Tajik AJ, Barnes ME, Ammash NM, Gertz MA, Seward JB, Oh JK. Differentiation of constrictive pericarditis from restrictive cardiomyopathy using mitral annular velocity by tissue Doppler echocardiography. *Am J Cardiol* 2004;94:316–319.
13. Shah D, Kumar CP, Shah MS, Baraiya M. Case series: congenital left ventricular diverticulum. *Indian J Radiol Imaging* 2010;20:211–214.