

Two cases of six-chambered hearts

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Double-chambered right ventricles (DCRV) and left ventricles are rare entities on their own. We present two cases with an unusual combination of double-chambered right as well as left ventricles. One was discovered in a 28-year-old female, while the other was found at birth in a female child. The differing nature of both the patient demographics as well as the presentation with a common morphological background is shown on both computed tomography and magnetic resonance imaging. The oldest description of an obstructive muscular band within the right ventricle was given in 1867. In the literature, there is ample description of the long-term prognosis and management guidelines for DCRV, but no inheritance patterns or risk factors have been identified except for associations with septal defects, tetralogy of Fallot and transposition of the great arteries. A combination of both double-sided left as well as right ventricles has been published in literature a few times with little details about management and prognosis.

SIMILAR CASES PUBLISHED: Although many cases of DCRV and DCLV have been published, to our knowledge only 3 cases of combined DCRV and DCLV have been published in literature.

Double-chambered ventricles are a rare congenital cardiac anomaly. The double-chambered right ventricle (DCRV) is seen much more frequently than the double-chambered left ventricle (DCLV).¹ However, a combination of the two is even rarer with fewer than ten case reports.² We present two cases with quadrichambered ventricular morphology in two very different patients with depiction of the cardiac anomaly on both magnetic resonance and computed tomography. No similar cases have been reported in literature.

CASE 1

A 28-year-old married female with a 1-year-old daughter presented with episodes of palpitations. There was no significant previous medical or surgical history with an uneventful pregnancy and normal delivery of a healthy child. General physical examination was unremarkable. Electrocardiograph failed to show any abnormality. An echocardiogram showed abnormal ventricular morphology described as a double-chambered right ventricle with a pressure gradient of 50 mm Hg and an hourglass constriction of the left ventricle with no pressure gradient. The pulmonary and aortic valves were normal. A cardiac MRI was advised for morphological assessment.

Cardiac MRI revealed situs solitus with normal atrial morphology and normal systemic and pulmonary venous drainage. However, both the right and left ventricles had horizontal bands that appeared to divide both the chambers in parallel. Ventricular contraction and myocardial thickness were normal and there was no abnormal enhancement pattern on post-gadolin-

ium delayed sequences. A subvalvular pulmonic gradient of 50 mm Hg was found on phase contrast imaging without abnormalities of the pulmonary valve. An ECG-gated cardiac CT was performed for further details. On CT images the ventricular bands appeared to be composed of myocardium as well as pericardium with insinuation of the pericardial fat into a groove forming a band-like structure that divided both the right and left ventricular cavities into a proximal and distal chamber. The band in the right ventricle was quite similar to a conventionally described double-chambered right ventricle morphology with origin of the pulmonary artery from the proximal chamber with consequent narrowing of the right ventricular outflow tract (**Figure 1**) however the band of the left ventricle did not cause narrowing of the left ventricular outflow tract (LVOT), nor was there any subaortic gradient (Video Clip 1 at <https://bit.ly/2JBWVXe>).

CASE 2

A female child of Kashmiri origin was born via normal vaginal delivery at 39 weeks of gestation with a normal cry at birth. There was no history of consanguinity or significant medical history in either parent. The mother was 29-years-old with an uneventful pregnancy

and an older male child aged 3 years with no significant medical history. Prenatal ultrasounds had shown normal development. The child was brought to a pediatrician for poor growth. Clinical examination was within normal limits and revealed no features of heart failure. However, there was a doubtful systolic murmur for which echo was advised. An echocardiography at 3 months of age revealed situs solitus with normal systemic and pulmonary venous drainage. The interatrial and interventricular septums were intact. Both tricuspid and mitral valves had normal morphology. The aortic valve was normal with normal great artery relationship and normal coronary arteries. There was biventricular hypertrophy with an infundibular/valvar pulmonary gradient of 50 mm Hg with a prominent muscle bundle in the right ventricular outflow tract. There was a suggestion of a thickened LV papillary muscle with laminar flow in the left ventricular outflow tract. For explanation of the biventricular hypertrophy and further elucidation of the DCRV morphology, a cardiac MRI was advised. The cardiac MRI at 4 months of age showed situs solitus with normal atrial morphology. Systemic and pulmonary venous connections were normal. Both the ventricles again in this case had abnormal muscular bands dividing each in parallel. Since there was normal systolic

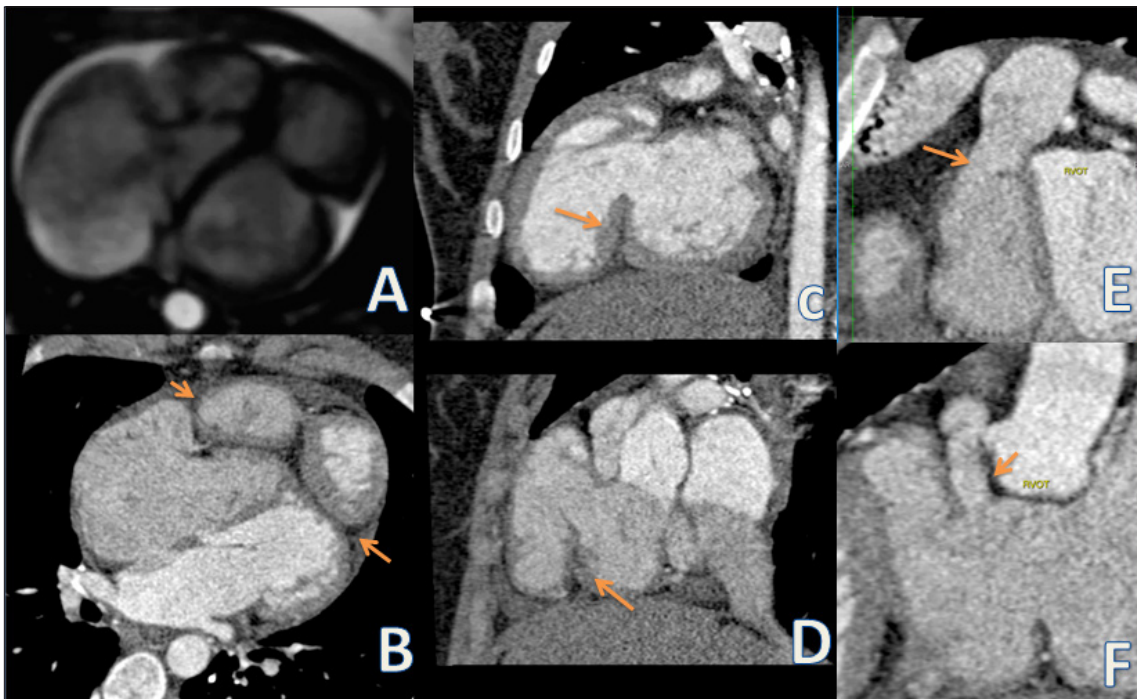


Figure 1. A) Cardiac MRI of four-chambered showing muscular bands in both the right and left ventricle. B) Cardiac-gated CT image showing band-like constriction around both ventricles with insinuation of pericardium. C and D) Sagittal reformatted views depicting the muscular bands in the right ventricle (C) and left ventricle (D). E and F) Sagittal and coronal reformatted views respectively depicting the narrowed right ventricular outflow tract.

thickening of the compacted myocardial layer, non-compaction was excluded. Also post gadolinium images revealed no delayed enhancement. Phase contrast imaging revealed a subpulmonic gradient of 50 mm Hg with a normal pulmonary valve. No subaortic or LV cavity gradient was noted. A cardiac CT was performed which again revealed a band-like constriction dividing the ventricles with insinuation of the pericardium along with the myocardium. The pulmonary artery arose from the proximal chamber of the right ventricle with a narrowed outflow tract (**Figure 2**) (Video Clip 2 at <https://bit.ly/2JBVVXe>).

DISCUSSION

A ventricle is rendered double-chambered by the presence of a whole or partial muscular ridge.¹ A double-chambered right ventricle is much more frequently reported than a double-chambered left ventricle. The oldest description of an obstructive muscular band within the right ventricle was given in 1867 by Peacock.¹ There is ample description of the long-term prognosis and management guidelines for DCRV, but no inheritance

patterns or risk factors have been previously identified for DCRV. However, associations have been established with septal defects, tetralogy of Fallot (TOF) and transposition of the great arteries.^{1,3,4} The DCRV has a low pressure distal chamber (anatomically higher) and a high pressure proximal chamber (anatomically lower).^{4,5} The chambers in DCRV are in series and cause eventual development of a gradient and a subinfundibular stenosis.^{3,4} The band is referred to as high when closer to the pulmonary valve and low when near the RV apex. This high pulmonary flow distal to the obstructing band differentiates DCRV from TOF. There is also systolic flattening of the ventricular septum with RV hypertrophy.⁴ DCRV can be caused by anomalous muscular bands or hypertrophied trabeculae.⁵ The right ventricle normally has a nonobstructing moderator band which may have a complex branching attachment. An aberrant moderator band also causes DCRV.⁴

DCLV has an unknown etiology with various theories considered like postinflammatory or hypoplasia of the myocardial wall or failure of regression of fetal trabeculations.³ Conventionally, seven subtypes of DCLV

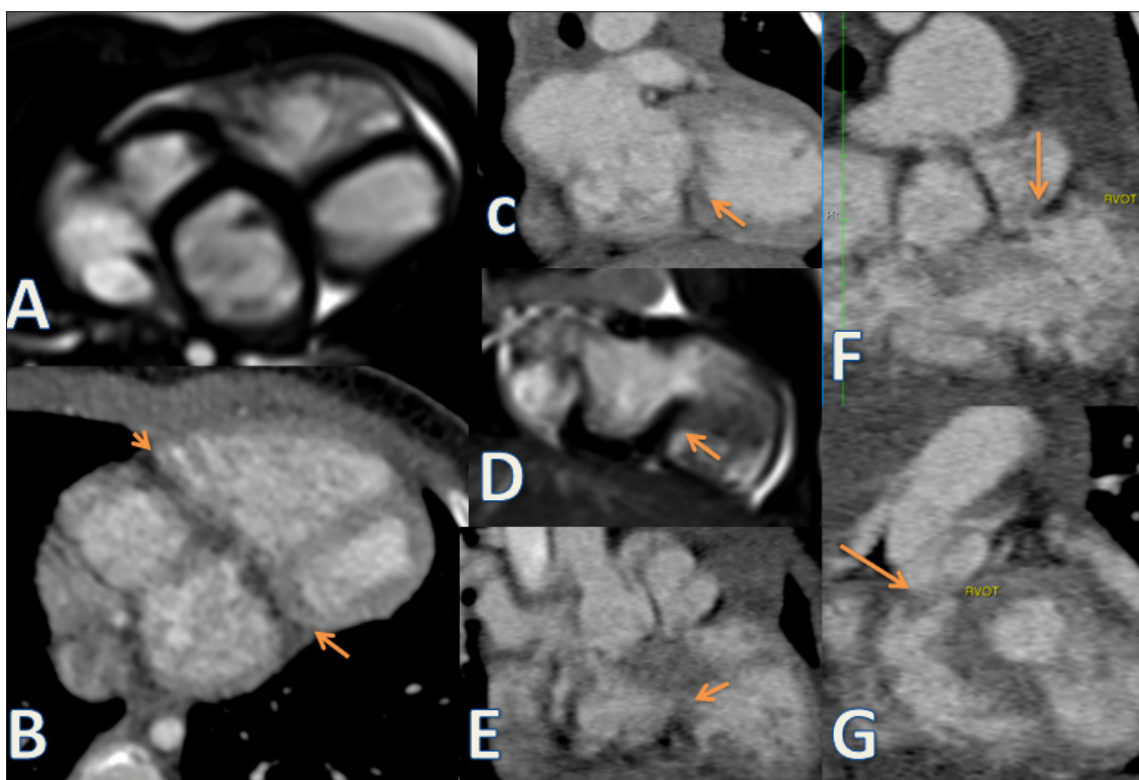


Figure 2. A) Cardiac MRI of four-chambered showing muscular bands in both the right and left ventricle. B) Cardiac-gated CT image showing band-like constriction around both ventricles with insinuation of pericardium. C, D and E) Sagittal reformatted views depicting the muscular bands in the right ventricle (E) and left ventricle (C) with coronal MRI depicting the LV band (D). F) Sagittal and coronal reformatted views depicting the narrowed right ventricular outflow tract.

had been described with four types as an aneurysm or diverticulum from the apex/base and three separate subtypes with differently positioned endocardial thickening. In one paper, the authors suggested defective development of myocardial intratrabecular sinusoids as causality.⁶ An intramyocardial aneurysm during embryonic life has been suspected as a possible event in the pathogenesis of DCLV.⁷ It has also been reported to be associated with endocardial fibroelastosis and mid-cavitary hypertrophic cardiomyopathy.⁴ An anomaly in the development of the papillary muscle from the myocardial ridges of the left ventricular walls has also been speculated as a cause of DCLV.⁸ The morphology of the left ventricle as such is divided into the "main" chamber and an "accessory" chamber. The size of the communication between the two determines the functional status with narrow access holes leading to possibly fatal ventricular dysfunction.⁷ The DCLV chambers are in parallel though and show synchronous contraction.³ Hence there is rarely flow acceleration of pressure gradient.⁴ An MRI finding of abnormally coarse and trabeculated left ventricular wall and a fragmented papillary muscles has also been described.⁸

While DCLV is most often detected incidentally, DCRV generally presents with murmur/dyspnea due to eventual development of a pressure gradient.³ Since the incidence of DCLV is quite low, ample data to determine prognosis and management is deficient.⁷ However, others have speculated that the management of DCLV may perhaps be guided by the degree of ventricular obstruction, the cardiac function of the patient and other associated anomalies that are often the primary concern when DCLV was identified incidentally.⁷

An important feature of double-chambered ventricles is the normal systolic contractility of the musculature. This allows differentiation from relatively common congenital diverticulae or aneurysms.³ However, aneurysms may have varying degrees of contractility depending on the amount of myocardial fibres involved.⁹ A DCLV band will not show late gadolinium enhancement, unless there is an underlying cardiomyopathy.^{4,8} Aberrant ventricular bands are another differential which can be present in both RV and LV and may contribute to LVOT obstruction. However, these are cord-like and fibromuscular and are differentiated from muscular

bands by their lack of contractility. These fibrous bands relax in systole and stretch in diastole while muscular bands become thick and short in systole.⁴ In both our cases the bands are muscular in nature. However, the peculiarity is the involvement of the pericardium along with the myocardium, which has not been described earlier. We found only three reports of a combination of DCRV and DCLV in the English literature.^{3,10,11} Since the embryological evolution of the DCLV and DCRV has been presumed to be separate in the literature, our cases raise the possibility of an association. The myocardium develops from cell lineages. The first heart field forms the left ventricular myocardium while the second heart field forms the right ventricle and the conotruncal region where influx of cardiac neural crest cells leads to the division of the aorta and pulmonary trunk.¹² With this background of distinct origins, it seems that the existence of a combined biventricular double chambers needs an embryonic event affecting both the first and second heart fields. At an embryonal stage, both the ventricles have a trabeculated appearance. In the LV these coalesce to form papillary muscles and most of the trabeculations disappear. Abnormal persistence of these trabeculations in the LV results in a non-compacted cardiomyopathy.¹³ A fibromuscular band dividing the LV, which on resection proved to be dense fibroelastic tissue, has been described.¹⁴ Ohlow described a partial stop in the development of the embryonic ventricle as a basis for formation of a congenital diverticulum or an in utero ischemic event.⁹ A focal persistence of trabeculations may cause a DCLV morphology by a hiatus during the embryonic phase as a fault in the endocardial-myocardial signaling.¹² Aberrant papillary muscles have also been described as cause of dynamic LVOT obstruction in hypertrophic cardiomyopathy.⁴ A superior-inferior LV subdivision by an abnormal papillary muscle has also been reported.⁸ Both our cases showed a "series" configuration of the DCLV chambers, with the pericardium accompanying the muscular band. Also, because of the large difference in the ages of our patients, a congenital etiology is probable. With an uncertain diagnosis and prognosis both these patients have been maintained on symptomatic treatment with regular follow up. Both have so far not reported any progression of symptoms or clinical deterioration.

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