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CASE SERIES
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A case series of double-chambered left ventricle detected by cardiovascular magnetic resonance

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Background Double-chambered left ventricle (DCLV) is a rare congenital condition, and few case reports are mentioned in literature. Entity, clinical course, and prognosis remain unclear. Cardiovascular magnetic resonance (CMR) is often used for characterization of various congenital heart diseases and can be particularly useful for imaging rare phenomena.

Case summary Three cases of DCLV were detected by CMR within 2 years in our CMR centre with and without associated congenital heart disease or hypertrabecularization. The patients did not suffer from cardiac symptoms despite the presence of premature ventricular complexes in one patient. Diagnosis of DCLV was made based on a first CMR study that was performed in adulthood, although some anatomical suspicion was already raised by previous echocardiography.

Discussion Double-chambered left ventricle, synonymous with the terminus 'cor triventriculare sinistrum', has been previously perceived as a rare phenomenon compared with double-chambered right ventricle. It has to be distinguished from ventricular aneurysm or cardiac diverticulum and is characterized by an additional contractile septum with normal wall structure that divides the LV cavum into two (rather) same-sized chambers. The prognosis seems to be benign, since there is no restriction in functionality and no increased thrombogenicity until adulthood. Consequently, there is (presumably) no need for a tailored therapy—at least in the cases present here. Accordingly, we recommend follow-up CMR examinations for progress monitoring and recognize CMR's significant role for diagnosis and follow-up of cardiac abnormalities in orphan diseases. Due to its broader availability, we expect further cases of DLVC in the future.

Keywords Double-chambered left ventricle • Double-chambered right ventricle • Cor triventriculare sinistrum • Cardiovascular magnetic resonance • Cardiac diverticulum • Case report

ESC Curriculum 2.3 Cardiac magnetic resonance • 9.7 Adult congenital heart disease

Learning points

- In case of suspected abnormalities of cardiac anatomy, a supplementary cardiovascular magnetic resonance should be performed despite an apparently good echocardiographic acoustic window.
- Double-chambered left ventricle seems to be not as rare as expected.
- Double-chambered left ventricle seems to have a good prognosis due to intact functionality and lack of thrombogenic milieu and remains often asymptomatic.

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Introduction

Cases of a double-chambered left ventricle (DCLV, synonymous with 'cor triventriculare sinistrum') were previously described only in a few case studies.[1] The configuration of a double-chambered ventricle occurs more often in the right ventricle (DCRV). Double-chambered right ventricle is a rare phenomenon as well, appears to be more complex, and is frequently associated with other congenital anomalies. In our cardiac magnetic resonance imaging (MRI) center, we identified three patients with the diagnosis of a DCLV over a 2-year period and within ~7000 MRI scans in total. As such, we believe that this cardiac abnormality is not as rare as expected, confirming the unique role of cardiovascular magnetic resonance (CMR) in the diagnosis of congenital heart disease compared with other imaging modalities.[2]

Timeline

Patient 1	At birth	Diagnosis of tetralogy of Fallot
	6 months	Correction surgery with ventricular septum defect (VSD) patch, partial resection of the pulmonary valve, transannular patch, and atrial septum defect (ASD) closure
	20 years	No cardiac symptoms First MRI and additional diagnosis of DCLV was made
Patient 2	Childhood	'Cardiac diverticulum' was detected
	37 years	No cardiac symptoms First MRI and diagnosis of DCLV was made
Patient 3	Childhood	'Left ventricular non-compaction' was detected
	About 55 years	Dizziness, headaches, and cardiac arrhythmias occur
	56 years	First MRI, no typical non-compaction, and diagnosis of DCLV was made

Patient 1

In the first patient, a 20-year-old woman, the diagnosis of the tetralogy of Fallot was made at birth and correction surgery was performed at 6 months including VSD patch, partial resection of the pulmonary valve, transannular patch, and ASD closure. In addition, an atypical origin of the left anterior descending coronary artery (LAD) from the right coronary artery (RCA) was known. She expressed no complaints and came to our hospital for routine check-up. There was no dyspnoea on exercise and no cardiac arrhythmia. The physical examination showed no abnormalities except for a 2/6 diastolic murmur in the second intercostal space. In the previous transthoracic echocardiography (TTE), the right ventricle (RV) was dilated, but with good function due to a higher degree pulmonary valve insufficiency. A double contour of the septum was seen and interpreted as a residuum of the VSD closure (*Figure 1*, patient 1). In addition to this, the subsequent CMR scan revealed an additional muscular septum that could neither be explained by the initial findings of a Fallot nor by the VSD patch surgery. Hence, the diagnosis of a 'DCLV' was made (*Figure 2*, patient 1). The complete patient characteristics and CMR findings are referred in *Table 1*. Due to lack of relevant symptoms, no specific therapy was initiated but a follow-up appointment was scheduled in our hospital in a 1-year period.

Patient 2

A 37-year-old woman was referred to us by a resident cardiologist. Her medical history comprised a 'cardiac diverticulum' and she presented for a routine check-up out of her own interest. She did not suffer from any cardiac complaints, and physical examination showed no abnormalities. Since echocardiography showed atypical diverticula (*Figure 1*, patient 2), we added CMR for an improved characterization. Cardiovascular magnetic resonance could not confirm a typical morphology of a diverticulum, but showed a muscular septation suggestive of a DCLV (*Figures 2* and *3*, patient 2). No therapy was necessary. We recommended further echocardiographic check-ups and electrocardiographies (ECGs) to detect potential asymptomatic arrhythmias.

Patient 3

A 56-year-old man, diagnosed with 'non-compaction cardiomyopathy' already at childhood, was complaining of dizziness, headaches, and cardiac arrhythmias for a year. Before that, he was free of any symptoms. He presented for the first time at our institution in good condition and without pathologies in the physical examination. Medical history revealed a mild form of left ventricular non-compaction in his sister, too. Echocardiography revealed some trabecularization in the left ventricle (LV), but no classic finding of spongy non-compaction (*Figure 1*, patient 3). The N-terminal prohormone brain natriuretic peptide (NT-proBNP) level showed a marginal increase of 292 pg/mL (normal value < 125 pg/mL). On resting ECG, we detected premature ventricular complexes. The CMR findings were not indicative of non-compaction cardiomyopathy, but an additional muscular septation with some trabecularization in the apical segments (*Figures 2* and *3*, patient 3). In addition, there was some additional trabecularization in the RV. In summary, we diagnosed a DCLV with atypical trabecularization. The patient received a ß-blocker (metoprolol succinate dose of 47.5 mg) for his arrhythmias and was scheduled for a follow-up visit in 1 year.

Discussion

Double-chambered left ventricle has previously been described as a rare phenomenon compared with DCRV. Although this condition is already considered a rare disease, it still has a higher incidence than DCLV. This can be explained best by the RV morphology. The RV is normally divided into an inlet and outlet by the crista supraventricularis, and pathologies of pre-existing anatomic regions are more common. A DCRV is more frequently associated with other congenital abnormalities, such as septal defects and/or tetralogy of Fallot. It is often symptomatic due to pressure gradients in the outflow tract caused by a progressive thickening of the right ventricular septum.[3,4] A few cases of DCLV or 'cor triventriculare sinistrum' have been previously published. It was rarely associated with other congenital diseases and was described as less symptomatic. Only very little reports exist on treatment and outcome.[1,5–9]

The double-chambered ventricle has to be distinguished from a congenital or acquired aneurysm, which is accompanied by a change in wall structure, that usually results in a hypo- to akinetic segments, and is histologically characterized by fibrous tissue with only isolated muscle fibres. Cardiac diverticula are also distinct from DCLV. The former is characterized by normal wall structure (endo-, myo-, and epicardium) and contractile tissue, while the opening to the cavum is often narrow and seen in 'appendix' morphology. In addition, diverticula are often associated with complex congenital heart defects and extracardiac malformations in the sense of midline thoraco-abdominal defects.[10,11]

In the cases described here, the muscular and normally contractile septum, which divides the LV cavum into two rather same-sized chambers, showed normal myocardial wall structure on CMR (*Figures 1* and *2*). Amongst others, the main advantage of CMR is non-invasive myocardial tissue characterization. By use of various T1- and T2-weighted

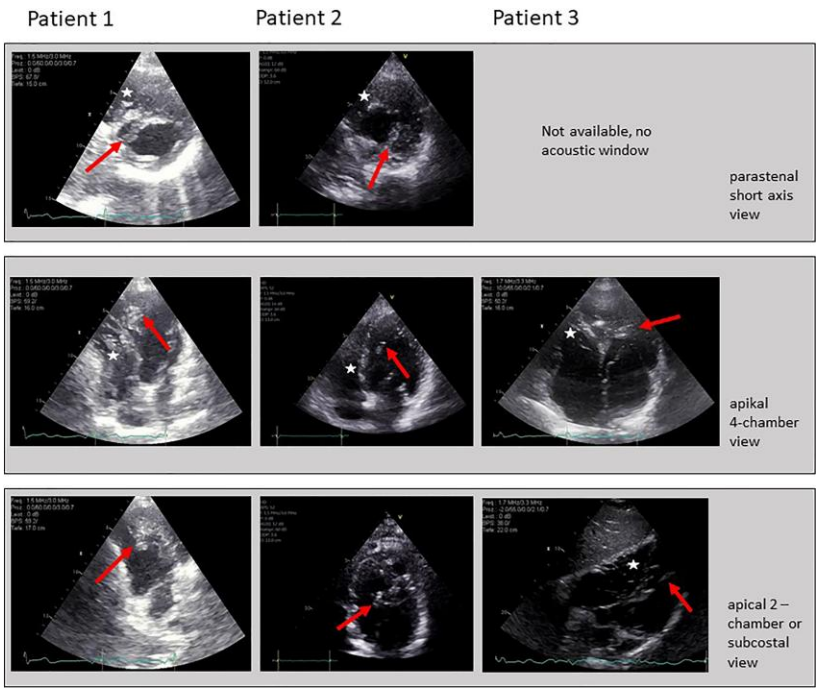

Figure 1 Presentation of double-chambered left ventricle in echocardiography. Arrow: additional left ventricle septum; Star: right ventricle cavum.

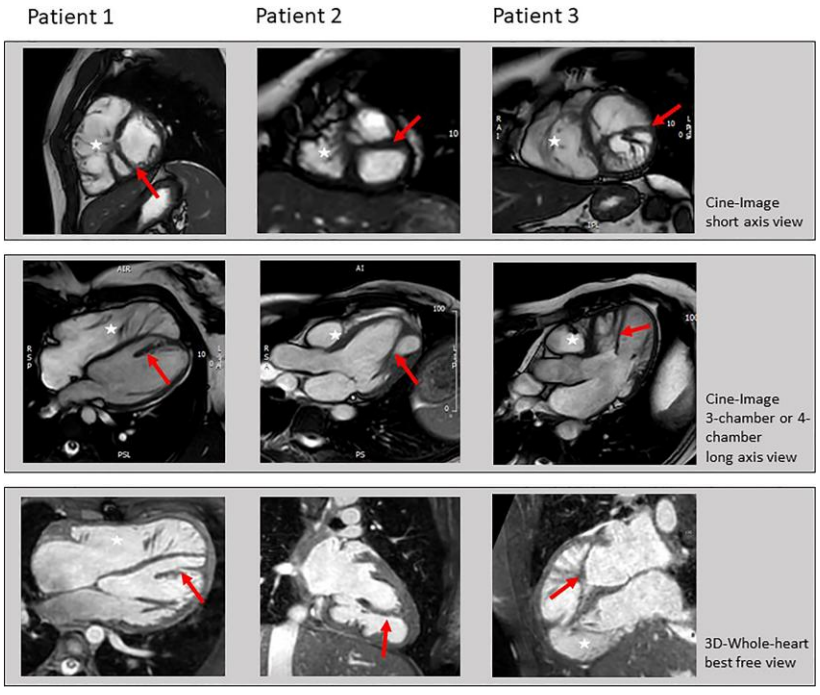

Figure 2 Presentation of double-chambered left ventricle in cardiovascular magnetic resonance—cine sequences. Arrow: additional left ventricle septum; Star: right ventricle cavum.

Table 1 Subject characteristics and CMR findings

	Patient 1	Patient 2	Patient 3
Sex	Female	Female	Male
Age at CMR, years	20	37	56
Diagnosis prior CMR	Tetralogy of Fallot	Left ventricular diverticulum	Non-compaction CMP
Symptoms	—	—	Premature ventricular complexes
Therapy	—	—	ß-Blockers
Additional septum	Septal to inferior wall Midventricular to apical	Septal to lateral wall Basal to apical	Septal to lateral wall Midventricular to apical
Associated malformations	Fallot formation and small muscular VSDs	—	—
Trabecularization	—	—	In the additional cavum
Contractility of the additional septum	Yes	Yes	Yes
LGE	not available	—	—
RV involvement	Enlargement because of Fallot	—	Trabecularization
LVEF, %	56	55	53
LVEDV, mL/m^2	79	87	85

CMR, cardiovascular magnetic resonance; CMP, cardiomyopathy; VSD, ventricular septum defect; LGE, late gadolinium enhancement; RV, right ventricle; LVEF, left ventricular ejection fraction; LVEDV, left ventricular end-diastolic volume

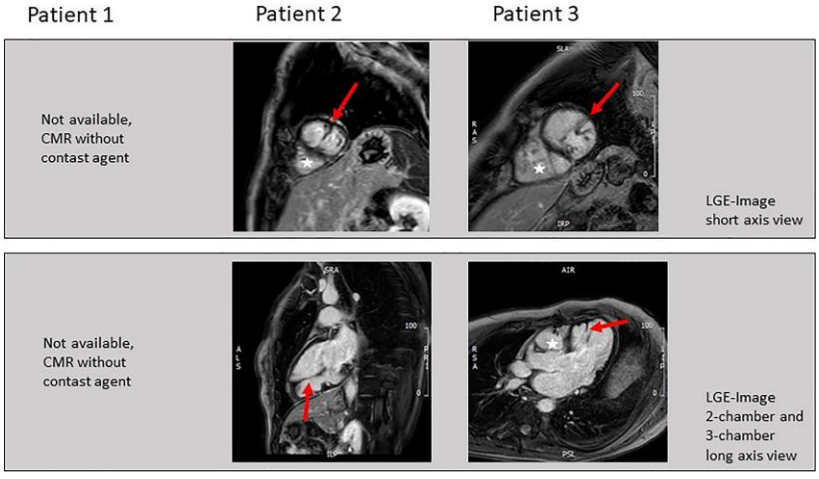

Figure 3 Presentation of double-chambered left ventricle in cardiovascular magnetic resonance—late gadolinium enhancement sequences. Arrow: additional left ventricle septum; Star: right ventricle cavum. CMR, cardiovascular magnetic resonance; LGE, late gadolinium enhancement.

sequences and late gadolinium enhancement imaging, fibrous or fatty tissues can be depicted and distinguished. No scar or otherwise abnormal tissue was found in our cases. Moreover, 3D imaging of the whole thorax, as performed in all patients presented here, enables the reconstruction of any arbitrary slice orientation in high-resolution (see Supplementary material online, *Figure S4*). Comparable results were not possible with echocardiography due to its limited thoracic window. The interplay and connection of these unique features helped us to clarify and refine a previously suspected echocardiographic diagnosis.

Some other characteristics need to be further addressed: First, in contrast to the configuration of a DCRV, no obstruction of the left ventricular outflow tract was found. This can probably be explained by the rather parallel arrangement of the chambers without any signs of different intracardiac pressure levels. Hence, no (regional) functional impairment and no abnormal muscular thickening were observed. Nevertheless, we recommend disease monitoring by clinical follow-ups at intervals of 1–3 years. Again, non-invasive CMR is particularly well suited in this regard, as it comes without almost no limits in penetration depth and can capture the entire cardiac structure and its function. Potentially progressive fibrosis can be depicted by late gadolinium enhancement images and parametric mapping techniques. In addition, flow measurements allow conclusions regarding pressure conditions. In the future, the wider use of new 4D flow sequences may provide even more detailed information about flow turbulence.

Second, regarding aneurysms, a tendency for thrombus formation is known due to the lack of kinetics and the changed wall properties. No thrombi have been seen so far in our cases and in the most cases described in the literature (only one case with coronary embolism).[12] Thus, it can be concluded that anticoagulation is not generally recommended, but its evaluation should always be an individual decision. Particularly for the detection of laminar or small thrombi, CMR offers unique advantages compared with other imaging methods.

Third, ventricular arrhythmias, which were also observed in a patient presented here, are described in the literature.[12] Whether ventricular arrhythmia is related to particularities of DCLV, or must be considered separately from it, is still unclear. Further, it is reasonable to assume that in context of a disturbed embryologic development, the conduction system may also be affected. Noteworthy, symptomatic bradycardia has not been mentioned so far in the available literature. We recommend Holter monitoring for detection of arrhythmia even in asymptomatic patients, because the arrhythmogenic risk is previously unknown.

Fourth, the trigger for this malformation, and whether it represents its own disease entity or is rather an epiphenomenon of other associated abnormalities, remains unclear. The frequently drawn connection to non-compaction CMP seems reasonably, and an atypical trabecularization was also present in one of our cases.[9] A strong link with other congenital diseases has not been described yet, although tetralogy of Fallot was present in one of our cases.

Conclusion

Through increasing availability of MRI and technical improvement of imaging modalities, anatomical peculiarities and congenital heart defects can be characterized earlier and better or more precisely. Consequently, conditions previously considered as rare are detected more frequently.

Double-chambered left ventricle or 'cor triventriculare sinistrum' seems to represent a rare anatomical variant with a good prognosis, since neither functional impairments nor increased thrombus formation was observed until adulthood. However, increased myocardial arrhythmia may be associated with this malformation and requires thorough monitoring. Therefore, we recommend regular Holter ECGs and follow-up examination by means of CMR, which is well suited for monitoring the progress of structural heart diseases, especially in those cases where echocardiography does not provide the desired image quality.

Lead author biography

Claudia Meier, born on 11 July 1983 in Germany, graduated from medical school at the University of Muenster, Germany, in 2010. She subsequently began and successfully completed her internal medicine fellowship with focus on cardiology in the University Hospital Muenster, Germany. She initially worked in the Department of Electrophysiology with focus on device therapy and began her specialization in cardiovascular magnetic imaging 2017 as a cardiologist under the guidance of Professor Ali Yilmaz, where she is the Deputy Head in the Division of Cardiovascular Imaging until now. She is an active member of the German Cardiac Society and European Society of Cardiology.

Supplementary material

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

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

Consent: The authors confirm that written patient consent for submission and publication of this case series including images has been obtained from the patient in line with COPE guidance.

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Data availability

The data underlying this article are available in the article and in its online supplementary material.

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