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HOW TO

How to look at adult congenital left ventricular outpouchings: a step-by-step approach using cardiac magnetic resonance

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Introduction

The definition 'congenital left ventricular (LV) outpouchings' describes any myocardial abnormality determining a protrusion of the LV cavity, often incidentally discovered during imaging tests like echocardiography, cardiac magnetic resonance imaging (cMRI) or cardiac computed tomography (cCT), usually performed for other reasons.

A knowledge of morpho-functional characteristics, embryology, and clinical implications is mandatory to interpret the radiological findings.

cMRI is one of the most used imaging tools for the evaluation of LV outpouchings, given its capability to assess morphology, localization, function, and tissue-characterization.[1] Recently, also cCT has reached a relevant position, comparable to CMRi, in this field, thanks to its spatial and temporal resolution.

The aim of the present '*how to*' article is to provide a brief step-by-step guide to approach the different outpouchings. We propose how to draft a comprehensive cMRI study protocol and interpret the results, making the differential diagnosis easier for imagers and clinicians.

Step 0: What are LV outpouchings

A uniform classification of LV outpouchings has not been established yet and their nomenclature is based on various imaging techniques and/or histopathological criteria, resulting in definitions not always consistent.[2]

'Crypts (clefts, fissures, or crevices)' are narrow invaginations within the myocardium,[1] composed of normal myocardial fibres, with systolic obliteration (explaining their rare post-mortem observation). They should penetrate >50% of the myocardial thickness; otherwise, they should be labelled as 'recesses'. They derive from a lack of compaction of trabeculated wall during embryogenesis. They are generally asymptomatic and free of prognostic significance, although been proposed as marker of disease in hypertrophic cardiomyopathy family members.[2]

'Congenital diverticula (muscular diverticula)' are 'finger-like' protrusions extending beyond the myocardial border (key difference with crypts[2]), connected to the main cavity throw a narrow neck.[2,3] They contain all the three ventricular layers (endocardium, myocardium, and epicardium) with preserved myocardial structure and minimal or absent fibrous tissue,[2,3] showing synchronous systolic contraction.

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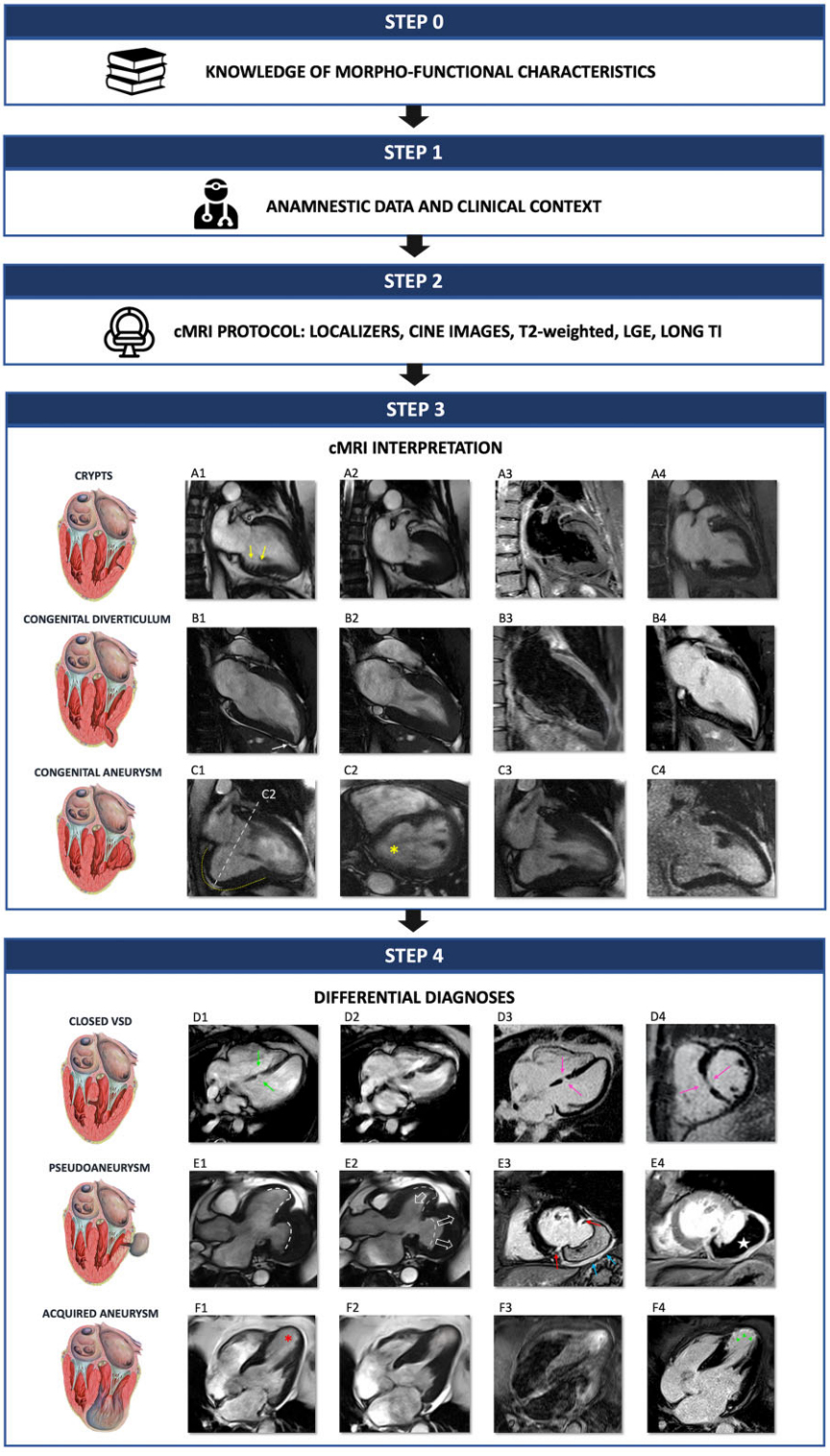

Figure 1 Step-by-step approach for the diagnosis of left ventricular outpouchings. Drawings modified from Patrick J. Lynch. Authorization details: Creative Commons Attribution 2.5 License 2006. cMRI, cardiac magnetic resonance imaging; LGE, late gadolinium enhancement; TI, time for inversion; VSD, ventricular septal defect.

Table 1 Main clinical, histopathological and morphological characteristics of congenital and acquired left ventricular outpouchings

	Different terms	Congenital vs. acquired	Clinical characteristics	Typical localization	Hystopathological characteristics	Morphological characteristics	Cine-sequences	Blood stasis on T2-w	LGE uptake
Crypts	Clefts, fissures, crevices; (recesses if <50% of the myocardium)	Congenital	Benign. Possible preclinical markers of HCM	Frequently basal inferior or septal wall	Normal myocardium	Generally multiple, narrow cavity. Penetrate >50% LV wall	Generally, no kinetic alteration, complete obliteration in systole	Absent	Absent
Muscular diverticula		Congenital	Generally asymptomatic. Possible adverse prognosis	LV apex	Normal myocardium. Possible fibrous tissue with thick pericardium	Thick wall, narrow neck. Finger-like, exceed LV wall	Generally, no kinetic alteration, obliterating in systole	Absent	Minimal or absent
Congenital aneurysms	Fibrous-type diverticula	Congenital	Rare but possible adverse prognosis	Sub-mitral or sub-aortic; LV apex	Normal myocardium. In LV apex possible isolated or atrophic myocardial fibres with connective tissue	Generally thin wall and wide neck; exceed LV wall	Akinetic or dyskinetic	Generally present	Generally present (lower in subvalvular forms). Normal surrounding myocardium
Spontaneous closed VSDs		Congenital	Generally asymptomatic	Septum	Fibrous tissue or myocardium	Small muscular defect with thin closure layer. Possible aneurysm	Kinetic alterations. Typically, not obliterating in systole	Possible	Possible, depending on the amount of fibrous tissue
Pseudo-aneurysms	False aneurysm	Acquired	Poor prognosis, variable risk of rupture, embolization or heart failure	Area of previous MI	Absence of myocardial layers with wall rupture plugged by pericardium or fibrous tissue. Surrounding myocardium involved	Discontinuity of LV wall. Thin layer, narrow neck	Dyskinetic	Present; possible thrombus	Present (possible also on surrounding myocardium)
Acquired aneurysms		Acquired	Poor prognosis, embolization or heart failure. Possible risk of rupture	Area of previous MI (mostly apical or anterolateral)	Fibrous tissue. Surrounding myocardium involved	Thin wall, wide neck; smooth transition from LV wall	Akinetic or dyskinetic	Present; possible thrombus	Present (also on surrounding myocardium)

'Congenital aneurysms' (sometimes misnamed double-chambered LV) are large thin-walled outpouchings composed of no or few working muscular fibres[3,4] (consequently akinetic or dyskinetic), usually connected to LV cavity through a wide neck.[3] Their location may be variable, though subvalvular (aortic or mitral) position is common. The aetiology of both congenital diverticula and aneurysms has been attributed to a focal embryogenetic defect of the ventricular wall.[3] Frequently asymptomatic, they may lead to adverse events (i.e., systemic embolization, valve regurgitation, heart failure, and arrhythmias).[3]

Step 1: Start with the clinical context

The differential diagnosis among congenital outpouchings and between them and acquired forms is challenging; a clinical and anamnestic evaluation is mandatory. Looking for other congenital abnormalities, systemic disease or syndromes, is fundamental to suspect specific entities. Careful exclusion of possible secondary causes (ischaemic, surgical, inflammatory such as sarcoidosis) is a key step in the differential diagnosis. The presence of symptoms is more consistent with acquired entities rather than with small diverticula or crypts.

Step 2: Cardiac magnetic protocol

To date, few and disaggregated data are available on cMRI capability in distinguishing LV outpouchings, without standardized and comprehensive criteria.[3] Every cMRI sequence gives us different but complementary information; here the most significant sequences in our experience.

Localizer scans

This set of low-resolution images can be the first to show the presence and localization of the outpouching, its relationship with other cardiac structures and extra-cardiac abnormalities possibly associated.

Cine sequences

Cine-images, performed with balanced steady-state-free-precession sequences (SSFPs), can be used in outpouchings examination for:

- Anatomic localization, using standard and off-axis projections.
- Morphological characteristics (wall thickness, extension through the wall layers).
- Functional characteristics (systo-diastolic motility but also biventricular function and wall motion abnormalities).

T2-weighted images

T2-weighted (T2w) black blood sequences can be used to evaluate blood stasis inside the outpouchings, since higher signal correlates with higher static blood content.

Postcontrast late gadolinium enhancement images

This technique has a crucial role in characterizing the composition of both the outpouching walls and the surrounding myocardium, detecting amount and distribution of late gadolinium enhancement (LGE) as marker of wall fibrosis. To better interpret these sequences, we recommend using diastolic frames instead of systolic ones, as shown in our examples. Post-contrast long 'time-from-inversion' (TI) sequences can detect thrombosis inside the outpouching cavity.

Step 3: Cardiac magnetic resonance interpretation

Crypts

- Localizer and SSFPs show narrow invaginations within the myocardium, perpendicular to the surface. Frequently multiples, their more common localization is the basal inferior wall[2] (*Figure 1*A1, yellow arrows). They show contraction and complete obliteration in systole, with normal kinetics of the surrounding myocardium (*Figure 1*A2).
- On T2w images (*Figure 1*A3), no signs of slow blood flow are visualized.
- On post-contrast images no signs of LGE are detected[1] (*Figure 1*A4).

Congenital diverticula

- Localizer and SSFPs show a 'finger-like' narrow-necked invagination[2,3] extending beyond the myocardial border (*Figure 1*B1, white arrow). LV apex is their most frequent location. They contract and almost obliterate in systole, without appreciable kinetic defects[2,4] (*Figure 1*B2).
- T2w images do not reveal signs of slow blood flow inside the diverticula (*Figure 1*B3).
- LGE sequences are useful to contour the shape of the outpouching, otherwise confoundable with the normal ventricular cavity; no signs of fibrosis are generally detected[1] (*Figure 1*B4).

Congenital aneurysms

- On localizer and SSFPs, they consist of a protrusion beyond the myocardial border, connected to the cavity through a wide neck[3] (*Figure 1*C1, yellow dotted line). They may be found in all LV regions, more typically in subvalvular location; their position and relationship are often better shown using off-axis projections (*Figure 1*C2, yellow asterisk). Given the paucity of normal-working myocardial fibres, localized akinesia is appreciable[2,3] (*Figure 1*C3).
- T2w images may show bright signal inside the cavity, due to slow blood flow.
- On post-contrast images, they usually show LGE;[2,3] adjacent myocardial regions are generally not involved (*Figure 1*C4). Of note, in agreement with other Authors, subvalvular aneurysms show minimal/absent fibrosis compared with those in other positions.[4]

Step 4: cardiac magnetic resonance clues for differential diagnosis

Closed ventricular septal defects

Spontaneous closure of ventricular septal defects (VSD) during intrauterine/postnatal period is frequent, with apposition of muscular or fibrous tissue, sometimes leading to aneurysm formation.[5] This

diagnosis should be considered when approaching septal outpouchings; anamnestic data are fundamental.

- On localizer and SSFPs, they appear as a thin-layer protuberance, located in the interventricular septum, sometimes protruding in the right cavity (*Figure 1*D1, green arrows). Cine-images show preserved contractility of the surrounding myocardium but without complete obliteration of the protrusion in systole[5] (*Figure 1*D2).
- T2w images can show bright signal due to local blood stasis.
- Post-contrast images can show LGE on the closed VSD layer, depending on the amount of fibrous tissue (*Figure 1*D3 and D4, pink arrows).

Acquired pseudoaneurysms

Pseudoaneurysms are acquired ventricular protrusions resulting from complicated MI or, rarely, surgery, trauma, or infection. They derive from a complete wall rupture, contained by thrombus, pericardium, or scar tissue, without a true myocardial layer.[1] A comprehensive assessment including clinical and instrumental data is fundamental.

- On localizer and SSFPs they appear as a pouch-like, narrow-necked sac (*Figure 1*E1). Cine-images show systolic dyskinesia of the pseudoaneurysm wall[1] (*Figure 1*E2, white arrows), extending to the surrounding myocardium.
- T2w images show intense bright signal inside the cavity (slow blood flow).
- On post-contrast images (*Figure 1*E3), it can be appreciable transmural LGE involving pseudoaneurysm neck (red arrows), partially extended to the adjacent ventricular wall and pericardium[1] (blue arrows). Frequently, voluminous thrombus can be found inside the cavity (*Figure 1*E4, white star).

Acquired aneurysms

Acquired aneurysms are extroversions of an akinetic wall, caused by fibrous replacement and thinning of the wall following transmural MI or other myocardial diseases (such as cardiac sarcoidosis). Their cMRI features are indistinguishable from congenital aneurysms; the differential diagnosis is based on clinical-anamnestic data, but also on the involvement of the surrounding myocardium, typically spared in the congenital forms.

- On localizer and SSFPs, they appear as large thin-walled LV protrusion with a wide neck; unlike pseudoaneurysms, the transition from the normal myocardium to the pathologic one is smooth, with no discontinuity[1,2] (*Figure 1*F1, red asterisk). The most common locations are apical or antero-lateral.[1] The local akinesia[1,3] (*Figure 1*F2) extends on a variable amount to the surrounding myocardium.
- T2w images show wide bright signal inside the aneurysm, due to blood stasis (*Figure 1*F3).
- On post-contrast images, transmural LGE, involving the surrounding LV regions, is appreciable[1] (*Figure 1*F4, green triangles).

Conclusions

Cardiac magnetic resonance has a crucial role in differential diagnosis of muscular wall abnormalities thanks to its capability in providing morphological and functional information as well as tissue characterization. A structured approach, as proposed, may be useful to examine and interpret cMRI images of LV outpouchings, never forgetting to integrate the radiological data with the clinical and anamnestic ones (*Table 1*). Finally, multimodality imaging approaches considering also cCT is useful for pre-procedural planning including the 3D-printing.

Conflict of interest: none declared.

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