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

Francesco Cardaioli, Giulia Brunetti, Alberto Cipriani, Raffaella Motta, Giuseppe Tarantini, Giovanni Di Salvo, Gaetano Thiene, Domenico Corrado, Cristina Basso, Sabino Iliceto, Manuel De Lazzari, and Martina Perazzolo Marra

1Cardiology Unit, Department of Cardiac, Thoracic, Vascular Sciences and Public Health, University of Padua Medical School, Padova, Italy; 2Department of Medicine, University of Padua, Padova, Italy; 3Pediatric Cardiology Unit, Department of Children and Woman’s Health, University of Padua, Padua, Italy; and 4Cardiovascular Pathology Unit, Department of Cardiac, Thoracic, Vascular Sciences and Public Health, University of Padua Medical School, Padova, Italy

Keywords: cardiac magnetic resonance • cardiovascular disease • congenital heart disease

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. 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.

‘Crypts (clefts, fissures, or crevices)’ are narrow invaginations within the myocardium, 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.

‘Congenital diverticula (muscular diverticula)’ are ‘finger-like’ protrusions extending beyond the myocardial border (key difference with crypts), connected to the main cavity throw a narrow neck. They contain all the three ventricular layers (endocardium, myocardium, and epicardium) with preserved myocardial structure and minimal or absent fibrous tissue, showing synchronous systolic contraction.
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 | Histopathological characteristics | Morphological characteristics | Cine-sequences | Blood stasis on T2-w | LGE uptake |
|-----------------|-------------------------|-------------------------|----------------------|-----------------------------------|--------------------------------|----------------|---------------------|------------|
| Crypts          | Congenital              | Benign. Possible preclinical markers of HCM | Frequently basal inferior or septal wall | Normal myocardium | Generally multiple, narrow cavity. Penetrate >50% LV wall | 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 | Absent | Minimal or absent |
| Congenital aneurysms | Fibrous-type diverticula | Rare but possible adverse prognosis | Sub-mitral or subaortic; 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 | Absent | Generally present |
| Spontaneous closed VSDs | Congenital | Generally asymptomatic | Septum | Fibrous tissue or myocardium | Small muscular defect with thin closure layer. Possible aneurysm | Possible | Possible, depending on the amount of fibrous tissue |
| Pseudo-aneurysms | False aneurysm | Acquired | 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 |
| Acquired aneurysms | Acquired | Poor prognosis, variable risk of rupture, embolization or heart failure | Area of previous MI (mostly apical or antero-lateral) | Fibrous tissue | Thin wall, wide neck; smooth transition from LV wall | Akinetic or dyskinetic | Present; possible thrombus |

**Congenital LV outpouchings:**

- Absent
- Minimal or absent
- Present (lower in sub-valvular forms)
- Possible, depending on the amount of fibrous tissue
- 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\(^1\)\(^,\)\(^2\)\(^,\)\(^3\)\(^,\)\(^4\)\(^,\)\(^5\) (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.\(^3\)\(^,\)\(^5\) 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\(^2\)\(^,\)\(^3\) (Figure 1A1, yellow arrows). They show contraction and complete obliteration in systole, with normal kinetics of the surrounding myocardium (Figure 1A2).
- On T2w images (Figure 1A3), no signs of slow blood flow are visualized.
- On post-contrast images no signs of LGE are detected\(^1\) (Figure 1A4).

**Congenital diverticula**

- Localizer and SSFPs show a ‘finger-like’ narrow-necked invagination\(^1\)\(^,\)\(^2\)\(^,\)\(^3\) extending beyond the myocardial border (Figure 1B1, white arrow). LV apex is their most frequent location. They contract and almost obliterate in systole, without appreciable kinetic defects\(^2\)\(^,\)\(^3\) (Figure 1B2).
- T2w images do not reveal signs of slow blood flow inside the diverticula (Figure 1B3).
- 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 1B4).

**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 1C1, 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 1C2, yellow asterisk). Given the paucity of normal-working myocardial fibres, localized akinisia is appreciable\(^2\)\(^,\)\(^3\) (Figure 1C3).
- 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 1C4). 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 1D1, green arrows). Cine-images show preserved contractility of the surrounding myocardium but without complete obliteration of the protrusion in systole (Figure 1D2).
- 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 1D3 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.  

- On localizer and SSFPs, they appear as a pouch-like, narrow-necked sac (Figure 1E1). Cine-images show systolic dyskinesia of the pseudoaneurysm wall (Figure 1E2, white arrows), extending to the surrounding myocardium.
- T2w images show intense bright signal inside the cavity (slow blood flow).
- On post-contrast images (Figure 1E3), it can be appreciable transmural LGE involving pseudoaneurysm neck (red arrows), partially extended to the adjacent ventricular wall and pericardium (blue arrows). Frequently, voluminous thrombus can be found inside the cavity (Figure 1E4, 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 (Figure 1F1, red asterisk). The most common locations are apical or antero-lateral. The local akinesia (Figure 1F2) extends on a variable amount to the surrounding myocardium.
- T2w images show wide bright signal inside the aneurysm, due to blood stasis (Figure 1F3).
- On post-contrast images, transmural LGE, involving the surrounding LV regions, is appreciable (Figure 1F4, 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.

**References**

1. Sharma A, Kumar S. Overview of left ventricular outpouchings on cardiac magnetic resonance imaging. Cardiowasc Diagn Ther 2015;5:464–70.
2. Cresti A, Cannarile P, Aldi E, Solan M, Sposato B, Franci L et al. Multimodality imaging and clinical significance of congenital ventricular outpouchings: recesses, diverticula, aneurysms, clefts, and crypts. J Cardiovasc Echogr 2018;28:9–17.
3. Ohlow M-A. Congenital left ventricular aneurysms and diverticula: definition, pathophysiology, clinical relevance and treatment. Cardiology 2006;106:63–72.
4. McMahon CJ, Moniotte S, Powell AJ, del Nido PJ, Geva T. Usefulness of magnetic resonance imaging evaluation of congenital left ventricular aneurysms. Am J Cardiol 2007;100:310–5.
5. Miyake T. A review of isolated muscular ventricular septal defect. World J Pediatr 2020;16:120–8.