Role of Cardiac Magnetic Resonance in the Diagnosis of Cardiac Amyloidosis

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Cardiac amyloidosis (CA) is characterized by progressive pathological deposition of amyloid fibrils in the myocardial interstitium, resulting ultimately in restrictive cardiomyopathy. The amyloid fibrils accumulating in the heart are most commonly misfolded immunoglobulin light chains (in case of AL amyloidosis) or transthyretin (ATTR), the latter being either in wild-type or variant conformation.

Overall, heart involvement is one of the strongest predictors of poor prognosis, with a median survival after diagnosis that ranges from less than 12 months for AL amyloidosis to 3–5 years for ATTR amyloidosis. Therefore, early detection and phenotyping of CA are crucial to stratify prognosis and guide treatment.

The pathological deposition of amyloid fibrils results in biventricular wall thickening and preserved ejection fraction (EF), at least until late. However, the systolic function of the heart is abnormal. Since the early stages of the disease, global longitudinal strain is characteristically reduced for both the ventricles, especially the basal segments, not following coronary artery distribution territories. Along with the progression of amyloid deposition, a transmural pattern of LGE can be seen. This is associated with a fivefold increase in mortality compared with patients with CA without LGE. In the most advanced stages, pan-myocardial LGE has been described (Fig. 1B).

Due to the high prevalence of severe chronic kidney disease in patients with systemic amyloidosis, a great interest has grown in native T1-mapping, which allows tissue characterization, without administration of contrast. Along with the expansion of the interstitial space due to amyloid accumulation, native T1-mapping and extracellular volume (ECV)-mapping are significantly elevated in patients with CA (Fig. 1C). This sign is very specific and correlates with prognosis. Moreover, elevated T1-mapping and ECV-
mapping could be found also in the absence or ill-defined LGE, thus acting as a potential early marker of disease.

In conclusion, CMR is fundamental for the diagnosis of CA. Its specificity and accuracy paid the way to the development of algorithms for a definitive diagnosis of CA without the need of biopsy. However, despite scores based on difference in the pattern of LGE have been proposed (QALE [quality adjusted life expectancy] score reference), it is still not possible to characterize the type of amyloid deposition based on CMR findings.

Therefore, clinical suspicion and expertise are needed to integrate the information derived from imaging into the clinical setting for the care of patients with CA.

Conflict of Interest
None.

References

Fig. 1 (A) SSFP four-chamber view showing left ventricular and left atrial hypertrophy. (B) Late-gadolinium enhancement distributed in both septum and atrium-ventricular walls. (C) T1-mapping native significantly elevated compatible with fibrosis. SSFP, steady state free precession.