



**Figure.** Example of fusion imaging with echocardiography (A) and computed tomography (B) during percutaneous occlusion of a postinfarction interventricular communication. Note the sheath passing through the defect.

phy,<sup>6</sup> found to be particularly useful in the study and closure of abnormal and multiple communications or residual defects after surgery.

Good results in structural catheterization can be obtained with precise monitoring of the procedure. In contrast, the use of a transesophageal probe for more than 60 minutes has been associated with an increase in oropharyngeal lesions, which may be indicated by an increased peripheral blood leukocyte count. A promising alternative is the transnasal probe, which, in addition to avoiding the need for general anesthesia, has shown good anatomical quality in various procedures. Fusion imaging (Figure) also safely reduces the procedural time and has become the best approach for transseptal puncture; for atrial appendage closure, fusion imaging with CT has also been shown to reduce the procedural contrast volume and time. Another emerging technology, especially suited to preprocedural planning, is 3-dimensional printing. Various articles have reported its ability to predict residual leaks after TAVI, test the safety of novel procedures, take accurate measurements and reduce the radiation dose in atrial appendage closure, and print material with different textures in the mitral valve field.

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## Selection of the Best of the Year 2017 in Cardiovascular Imaging in Familial Cardiopathies

### Selección de lo mejor del año 2017 en imagen cardiovascular en las cardiopatías familiares

To the Editor,

The use of cardiovascular imaging techniques (CVIT) in clinical processes in the context of inherited cardiac conditions (ICC)



presents specific opportunities and challenges in a clinical scenario marked by ever-changing understanding of their biology, the speed of technological development, and the need to adapt to the different social, economic, and health characteristics of each population.

The availability of different CVITs for approaching the same clinical problem can make it difficult to choose the most appropriate technique. A consensus document from several North American societies aims to present, in a structured way, the appropriate use criteria for the various CVITs in primary and secondary valvular heart disease in patients with ICC.<sup>1</sup> However,

the real essence of the multimodal CVIT approach consists of capitalizing on the most robust features of each technique so that they complement each other, which allows identification of the aspects of a clinical condition that other imaging modalities cannot.

Arrhythmogenic cardiomyopathy is a disease with progressive morphofunctional expression, and various CVITs are usually used periodically throughout the clinical course of the disease. It therefore serves as an example for a systematic multimodal approach, and was featured in a recent European document that makes for recommended reading,<sup>2</sup> in which the authors also elaborate on some of the diagnostic criteria published in 2010.

The clinical teams in charge of ICC often receive patients with atypical cardiomyopathies awaiting diagnosis; they also frequently see cases in which the origin is not genetic, as presumed, but autoimmune or autoinflammatory. An excellent summary has recently been published on systemic immune-mediated diseases, and cardiologists should be aware of its interesting practical recommendations.<sup>3</sup>

However, one of the most important current aims for research on CVITs in ICC is undoubtedly to demonstrate their role in real clinical populations and test their effective prognostic usefulness. In a descriptive multicenter study, 852 consecutive patients with bicuspid aortic valve were evaluated with transthoracic echocardiography.<sup>4</sup> The most prevalent morphotype was coronary cusp fusion (72.9%), followed by fusion of the right coronary and noncoronary cusps (24.1%), and the rarest morphotype was that resulting from fusion of left coronary and noncoronary cusps (3.0%). Bicuspid aortic valves without raphe represented less than one fifth of the total. Dilatation of the sinuses of Valsalva was present in 34% of cases, and was associated with male sex and aortic regurgitation. In addition, the second morphotype (right coronary–noncoronary fusion) was found to be a protective factor against aortic root dilatation. However, dilatation of the tubular ascending aorta was more prevalent (76%) and was present in equal amounts in the different valvular morphotypes.

Prediction of sudden death in cardiomyopathy is essential because it may be the first clinical manifestation of the disease, but use of implantable cardioverter-defibrillators as primary prevention in very low risk populations has unacceptable clinical and financial costs. Prognostic assessment in ICC is complicated. There are several risk markers in the various conditions, but it is difficult to translate these to clinical decision making because, among other things, the relationship between the severity of the risk marker and its predictive value can be complex and nonlinear. For example, in a retrospective multinational cohort study that examined the clinical outcomes of 3673 patients with hypertrophic cardiomyopathy after 5 years of follow-up, the investigators established that the risk of sudden death in patients with a maximal parietal wall thickness  $\leq 14$  mm was comparable to that of patients with maximal parietal wall thickness  $\geq 35$  mm; therefore, it is inappropriate to assume a linear relationship between myocardial thickness and sudden death, and the decision to implant a defibrillator should not be based on the presence of extreme ventricular hypertrophy alone.<sup>5</sup> In another study of patients with

nonischemic dilated cardiomyopathy without severe systolic dysfunction, the presence of late myocardial gadolinium enhancement on magnetic resonance imaging was associated with an increased risk of sudden death, but the relationship between the extent of enhancing myocardium and the degree of risk was not clearly linear.<sup>6</sup> It is expected that CVITs will become well-established as diagnostic and prognostic tools for ICC in everyday clinical practice.

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