dilated by PTPA. Meanwhile, lesion type B is moderately difficult to identify by angiography, and OCT revealed a honeycomb-like structure. Lesion type C is a representative lesion that is difficult to identify by angiography. OCT revealed a lotus root-like structure in the pulmonary artery in this lesion before PTPA, and impaired flow was confirmed by pressure ratio. Blood flow could pass through some of the spaces in the lotus root-like structure, but each of these flow spaces was significantly narrowed, which may explain why this lesion was difficult to identify by angiography and had a significantly reduced pressure ratio. OCT after PTPA demonstrated that the lotus root-like structure was destroyed by balloon dilation and crushed toward the vessel wall, accompanied by significant improvement of pressure ratio (>0.8), demonstrating that the lesions with lotus root-like structures can be easily dilated by PTPA. Lesion type D, located within a pulmonary artery that has unimpaired blood flow and with a pressure ratio that was detected by pressure wire, was also within the normal range. However, OCT demonstrated a lotus root-like structure within this vessel, with possible organized thrombus adherence to the vessel wall. The main space through which blood flow passed was not significantly narrowed, which may explain why this vessel had a lotus root-like structure but no impaired flow. Interestingly, OCT in lesion type D demonstrated that normal angiographic findings and a preserved value of pressure ratio do not necessarily ensure normal structure of vessels in CTEPH. These findings suggested that the lotus root-like structure could be seen in 2 cases (i.e., lesions with or without impaired flow) but both are difficult to identify by angiography.

PTPA has recently been developed and needs further improvement. The findings in this report suggest that OCT is a useful tool to demonstrate the morphological features of lesions. In particular, OCT is useful to detect target lesions that are difficult to identify by angiography. A combined approach, using angiography as a subjective tool to evaluate flow grade, a pressure wire to provide an objective value of flow impairment, and OCT as a useful tool to demonstrate the concrete morphological features of a lesion, may enable more appropriate judgment regarding the PTPA strategy.

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http://dx.doi.org/10.1016/j.jcmg.2015.02.015

Please note: The authors have reported that they have no relationships relevant to the contents of this paper to disclose. The first 2 authors contributed equally to this work.

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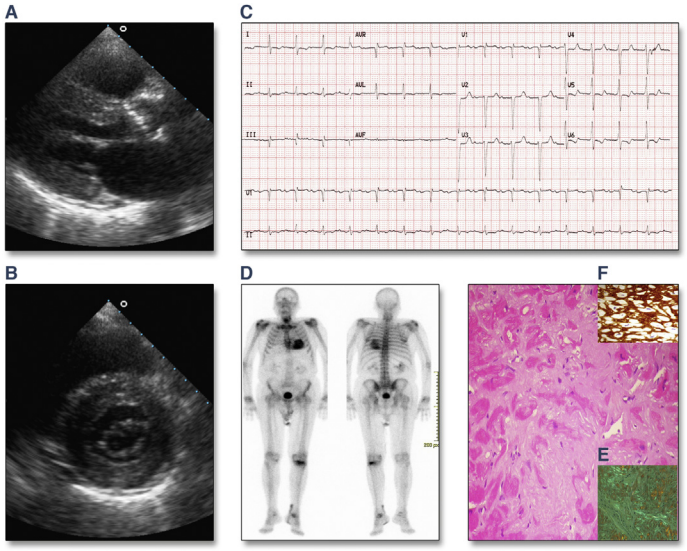
Coexistence of Degenerative Aortic Stenosis and Wild-Type Transthyretin-Related Cardiac Amyloidosis

Degenerative aortic stenosis (AS) is a growing cause of heart failure and death in the elderly. The majority of patients with symptomatic AS are currently treated with surgical or transcatheter aortic valve replacement (TAVR). It was recently suggested (1) that a share of the sometimes lethal complications arising during and after aortic valve replacement are to be attributed to coexistent cardiac amyloidosis. In an autopsy series of patients who had undergone TAVR, Nietlispach et al. (2) found varying degrees of ventricular myocardium amyloid infiltration in approximately one-third of cases. The etiology of amyloidosis was not defined, but considering patient age and clinical profile, it is plausible that these patients had wild-type transthyretin

JACC: CARDIOVASCULAR IMAGING, VOL. 9, NO. 3, 2016
MARCH 2016:321–31

FIGURE 1 Patient With Aortic Stenosis and Wild-Type TTR-Related Amyloidosis



(A and B) Echocardiogram showing "red flags" for cardiac amyloidosis: ventricular myocardium granular sparkling, atrioventricular valve thickening, and mild pericardial effusion. **(C)**. Electrocardiogram: typical signs of severe aortic stenosis (left ventricular hypertrophy) are absent. **(D)** Technetium-99m-3,3-diphosphono-1,2 propanodicarboxylic acid scintigraphy showing strong myocardial tracer uptake. **(E and F)** Endomyocardial biopsy with TTR-related amyloid infiltration. **(E)** Hematoxylin-eosin, Congo red staining. **(F)** Immunohistochemistry with anti-TTR antibodies. TTR = transthyretin.

(TTR)-related amyloidosis (wt-ATTR). Indeed, degenerative AS and wt-ATTR share a common demographic and clinical profile. Data from postmortem studies in unselected subjects indicate a prevalence of cardiac amyloidosis of 22% to 25% in subjects older than 80 years of age (3).

TTR-related cardiac amyloidosis can currently be accurately identified by scintigraphy with bone-seeking tracers (4). A recent study with technetium-99m-3,3-diphosphono-1,2 propanodicarboxylic acid (^{99m}Tc-DPD) scintigraphy in 12,400 subjects found a prevalence of strong, unexpected myocardial tracer uptake, suggestive of cardiac amyloidosis, in ~1.4% of subjects older than 80 years of age (5).

To clinically investigate the coexistence of cardiac amyloidosis in elderly patients with AS referred for aortic valve replacement (surgical or transcatheter), we prospectively evaluated with ^{99m}Tc-DPD scintigraphy all patients with degenerative AS and 1 or more of the following echocardiographic "red flags": increased thickness of atrioventricular valves, interatrial septum or right ventricular free wall, pericardial effusion, and myocardial granular sparkling.

Of the 43 patients referred with AS (between October 2014 and January 2015), 5 had echocardiographic "red flags" and underwent ^{99m}Tc-DPD scintigraphy before a final decision regarding treatment was made. The scan showed strong myocardial uptake in all patients. All then underwent endomyocardial biopsy, which consistently demonstrated TTR-related amyloid infiltration. Genetic analysis excluded TTR gene mutations, and wt-ATTR was diagnosed in all 5 patients.

Of the 5 patients with wt-ATTR, only 1 was female (median age 84 years; range 76 to 90 years), and all had advanced heart failure (New York Heart Association functional class III/IV). Interestingly, 3 had a history of carpal tunnel syndrome (bilateral in 2 patients). All had marked symmetrical left ventricular (LV) hypertrophy (median wall thickness, 18 mm; range 16 to 21 mm) with a calculated aortic valve area ≤0.6 cm^2/m^2. In 4 of 5 patients, a low flow-low gradient condition was present (with reduced LV ejection fraction in 2 patients). Remarkably, all echocardiographic "red flags" were present in 3 of 5 patients, and longitudinal function was depressed in all patients. A disproportion between echocardiographic LV "hypertrophy" and normal QRS voltages was present in 4 patients (only 1 satisfied electrocardiographic criteria for LV hypertrophy). All patients underwent balloon aortic valvuloplasty with a "bridge to decision" strategy and are currently undergoing a clinical follow-up. Figure 1 summarizes the instrumental findings of a typical case.

This small preliminary study found that the coexistence of degenerative AS and wt-ATTR (a potentially dangerous condition in patients undergoing aortic valve replacement) can be suspected on the basis of clinical and echocardiographic elements and can be effectively diagnosed by ^{99m}Tc-DPD scintigraphy. The study cannot provide data on disease prevalence and test sensitivity but suggests that particular attention should be paid to patients with echocardiographic signs of myocardial infiltration and/or a mismatch between LV wall thickness and QRS voltages and/or low flow-low gradient and depressed longitudinal myocardial function. Larger prospective studies are needed to establish in which patients with AS to search for cardiac amyloidosis as well as how to use this information in the treatment decision-making process.

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http://dx.doi.org/10.1016/j.jcmg.2015.04.012

Please note: This study was partially supported by the strategic program "Regione Emilia Romagna–Università 2010-2012 Technological Innovations in the Treatment of Heart Failure" and by an unconditioned Research grant from Pfizer Inc. Pfizer had no role in the study design, data analysis, and results interpretation of the present study. Dr. Saia has received consulting fees from Abbott Vascular, Eli Lilly, AstraZeneca, and St. Jude Medical; and speaker fees from Abbott Vascular, Eli Lilly, AstraZeneca, St. Jude Medical, Terumo, Biosensors, Edwards Lifesciences, and Boston Scientific. Dr. Rapezzi has received consulting and speaker fees from Pfizer Inc. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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How to Distinguish Between Myocarditis and Sarcoidosis Based on CMR Imaging

I read with great interest the manuscript "Late Gadolinium Enhancement Among Survivors of Sudden Cardiac Arrest" (1). This paper offers the best available guidance on the management of this complex group of patients, which we recently utilized in the management of one of the patients admitted to our hospital. As someone who performs cardiac magnetic resonance imaging, I am often confronted by the inability to distinguish between active myocarditis and cardiac sarcoidosis, especially when the ventricles do not exhibit gross structural abnormalities. I find the delayed gadolinium enhancement pattern alone insufficient in distinguishing between these conditions unless extra-cardiac sarcoidosis is manifest.

Perhaps this distinction is moot since late gadolinium enhancement turns out to be the single best predictor of future appropriate implantable cardioverter-defibrillator therapy. On the other hand, this may be important because treatment for sarcoidosis may be available. Additionally, a clear definition of the mechanisms/disease that leads to sudden cardiac death is important from the epidemiologic point of view. Therefore, it would be helpful for the investigators to highlight the methods utilized to distinguish between these conditions.

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http://dx.doi.org/10.1016/j.jcmg.2015.05.015

Please note: Dr. Aneja has reported that he has no relationships relevant to the contents of this paper to disclose.

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THE AUTHORS REPLY:

We thank Dr. Aneja for the interest in our article (1) and also appreciate that the feedback that the information provided in the article was of use when you were involved in the care of a patient surviving sudden cardiac arrest.

Sarcoidosis consists of both an active, inflammatory phase as well as a chronic phase, in which scarring and fibrosis are present. Although the cardiac magnetic resonance (CMR) features of the 2 conditions overlap especially during the acute phase, there are some suggestive features that help differentiate the two (2,3). The following features are typically seen in cardiac sarcoidosis (2,3):

1. Late gadolinium enhancement (LGE) in cardiac sarcoidosis often involves more than one segment, in a basal distribution, and often involves the septum.