

pro-brain natriuretic peptide (NT-proBNP) (4858.0 [3047–5801] pg/mL vs 3407.0 [2188–4853] pg/mL;  $P = .3$ ). Similarly, there were no differences between baseline and end of follow-up glomerular filtration rate (45.9 [36.9–59.7] mL/min/1.73m<sup>2</sup> vs 47.0 [43.6–105.0] mL/min/1.73m<sup>2</sup>;  $P = .3$ ).

This study examined a cohort of patients with aHF who received CIL as a bridge to heart transplant. Follow-up was longer than in previous reports,<sup>1,2</sup> and ICDs were interrogated periodically, allowing analysis of levosimendan safety in patients included on a HTWL. Only 22% of the patients required an emergency heart transplant, contrasting with emergency transplant rates of 64% and 44% for HTWL patients in European and Spanish registries, respectively, in 2017.<sup>5,6</sup> These data indicate that CIL is a practical bridge-to-transplant option.

Levosimendan infusion was safe in all patients, with no incidents of ventricular arrhythmia recorded during treatment or follow-up; however, the sample size is too small to allow definitive conclusions. Nevertheless, our results are important, since the prolonged action of the drug means that beneficial and adverse effects will not be limited to the infusion, but will also manifest in the days afterwards. The most concerning adverse effects are ventricular arrhythmias, but to our knowledge, no previous study has analyzed the occurrence of arrhythmias in the postinfusion period. Moreover, the heart failure admission rate in our cohort was lower than that reported in previous studies.<sup>1,2</sup>

## FUNDING

This study was supported by the *Instituto de Salud Carlos III* and the European Regional Development Fund (ERDF) through the *Centro de Investigación Biomédica en Red de Enfermedades Cardiovasculares* (CIBERCV).

## CONFLICTS OF INTEREST

Juan F. Delgado has delivered presentations at Orion Pharma conferences and has participated in clinical trials funded by Orion Pharma. Javier de Juan and Inés Ponz have delivered presentations at Orion Pharma conferences.

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Available online 13 February 2020

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<https://doi.org/10.1016/j.rec.2019.10.026>  
1885-5857/

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## Arrhythmogenic right ventricular cardiomyopathy presenting as myocarditis in young patients: a concealed relationship



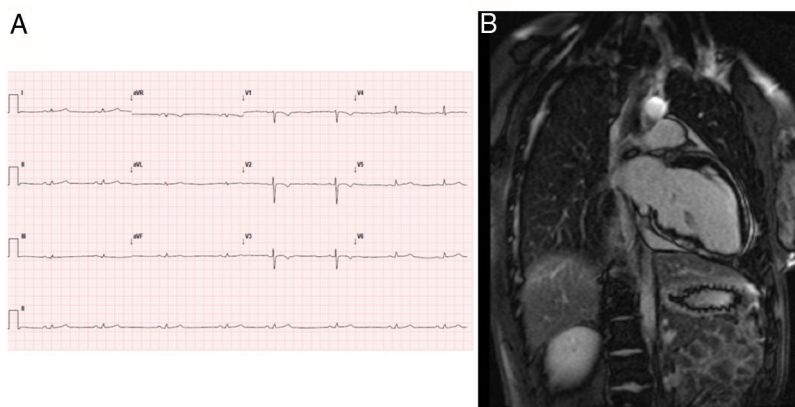
### Miocardopatía arritmogénica del ventrículo derecho en pacientes jóvenes con miocarditis: una asociación oculta

#### To the Editor,

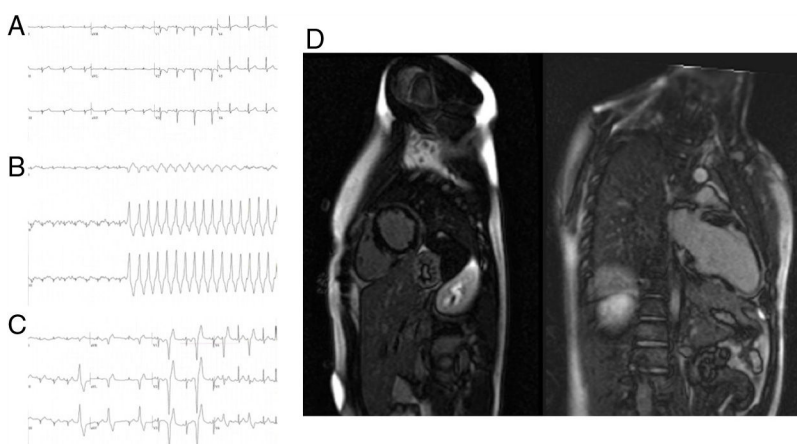
Diagnosis of arrhythmogenic right ventricular cardiomyopathy (ARVC) can be challenging. Recent evidence indicates that the natural history of this disease includes a first concealed phase, characterized by acute exacerbations of myocardial inflammation and life-threatening ventricular arrhythmias, occurring prior to the onset of classical characteristics and contributing to its pathogenesis and progression.<sup>1</sup> This has been demonstrated by reports of ARVC presenting as recurrent myocarditis-like episodes in young patients with evidence of myocardial inflammation on cardiac magnetic resonance.<sup>2</sup> Instead of the classical replacement in this disease of myocytes by fibrous or fibroadipose tissue in the right ventricular (RV) myocardium,<sup>3</sup> inflammatory infiltrates can often be seen in affected areas.<sup>4</sup> This

article intends to illustrate this association, making a compelling argument for a thorough investigation of the RV in young patients presenting with ventricular arrhythmias and signs of active or past myocarditis.

Patient 1, a previously healthy girl, presented at the age of 15 years with aborted sudden cardiac death during competitive sports. Rhythm was pulseless ventricular tachycardia. The baseline electrocardiogram (ECG) showed low voltage and T wave inversion in the right leads (figure 1), thought to be normal for her age. Her father had the same T wave pattern, but family history was otherwise not relevant. One week before the current event, she was diagnosed with tracheobronchitis, with 1 day of fever. During the current admission, she progressed well. Twenty-four hour Holter monitoring showed isolated, polymorphic ventricular ectopic beats (28 beats/h). Cardiac magnetic resonance revealed indirect signs of active inflammation with enhanced spontaneous left ventricular (LV) myocardial SSFP signal, and multiple locations of subepicardial late gadolinium enhancement (LGE), which were more evident at the inferior LV wall (figure 1). LV ejection fraction (EF) was 58%, and end-diastolic volume was normal (85 mL/m<sup>2</sup>). RV ejection fraction was 48%, and end-diastolic volume was at the upper



**Figure 1.** Patient 1. A: ECG with low voltage and T wave inversion at the right precordial leads. B: cardiac magnetic resonance with subepicardial late gadolinium enhancement, especially at the basal and mid-inferior left ventricular wall.



**Figure 2.** Patient 2. A: baseline ECG with T wave inversion at the right precordial leads. B: ventricular tachycardia during exercise testing with inferior axis. C: ventricular ectopic beats with RVOT origin. D: cardiac magnetic resonance showing extensive subepicardial late gadolinium enhancement, especially at the inferolateral left ventricular wall.

limit of normal ( $108 \text{ mL/m}^2$ ). Mild dyskinesia of the right ventricular outflow tract (RVOT) was noted ([video 1 of the supplementary data](#)), but was not consensual. At the time, a presumptive diagnosis of acute myocarditis was made, despite negative etiologic investigation. An implantable cardioverter-defibrillator (ICD) was implanted. Genetic screening by a sequencing method revealed a previously unknown variant in heterozygosity: c.1840delC in the *PKP2* gene, causing protein truncation, diagnostic for ARVC. The father is a gene carrier, exhibiting only the ECG abnormalities. His extended family was not studied. The patient has been followed-up and has been well for 2 years.

Patient 2, a 13-year-old boy, was referred due to ventricular ectopia of left bundle branch block morphology in the last 2-yearly ECGs performed for competitive sports practice. ECGs also showed T wave inversion in the right precordial leads ([figure 2](#)), considered normal for his age. The family history was negative and the patient was previously healthy except for an episode of oppressive chest pain 2 years earlier lasting a few days and preceding the appearance of ectopia. Upon current evaluation, he mentioned light-headedness and palpitations during sports. The echocardiogram revealed mild RV dilation. Exercise testing elicited non-sustained RVOT ventricular tachycardia ([figure 2](#)) and multiple isolated RVOT ventricular ectopic beats ([figure 2](#)). Cardiac magnetic resonance showed normal LV dimension and function,

but extensive subepicardial LGE ([figure 2](#)). The RV was mildly dilated ( $115 \text{ mL/m}^2$ ), and EF was 39%, with mild apex and RVOT dyskinesia ([video 2 of the supplementary data](#)). Due to the patient's previous history of chest pain, LGE pattern and RV dyskinesia, doubts were elicited about the correct diagnosis: scar tissue from a previous episode of myocarditis and/or ARVC manifested by a previous inflammatory exacerbation. Beta-blocker therapy was instituted. While awaiting implantable cardioverter-defibrillator implantation, the patient experienced an episode of sustained RVOT tachycardia. Follow-up electrophysiologic study induced and successfully ablated this tachycardia. Another form of faster ventricular tachycardia originating from the apex was induced but not sustained sufficiently to allow mapping. A subcutaneous implantable cardioverter-defibrillator was implanted. High resolution signal-averaged ECG showed late potentials. Genetic testing by a sequencing method revealed a previously undescribed variant in heterozygosity: c.224-2T>C in the *PKP2* gene, leading to protein truncation and diagnosis of ARVC. First-degree relatives declined study. The patient has been medicated with sotalol, followed-up, and has been asymptomatic for 1 year.

These cases illustrate how ARVC can present early in life with life-threatening ventricular arrhythmias and inferolateral subepicardial LGE. This is a common LGE pattern both in myocarditis<sup>5</sup> and ARVC with LV involvement.<sup>1</sup> Therefore, myocar-

ditis, which is more prevalent at such a young age, may be precipitously diagnosed. Careful consideration of other diagnostic cues, such as RV size and regional contractility in our patients, may lead to comprehensive genetic testing<sup>6</sup> and uncover the underlying disease. In fact, both patients met task force criteria<sup>3</sup> (3 major + 1 minor and 2 major + 2 minor criteria, respectively). Achieving a correct diagnosis of ARVC has major implications for future management and familial risk stratification.

## APPENDIX. SUPPLEMENTARY DATA

Supplementary data associated with this article can be found in the online version, at <https://doi.org/10.1016/j.rec.2019.10.007>

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Available online 18 November 2019

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<https://doi.org/10.1016/j.rec.2019.10.007>  
1885-5857/

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## MSCT-fluoroscopy fusion imaging for transcaval access guidance in transcatheter aortic valve replacement



### Imagen de fusión TCMC-fluoroscopia en el reemplazo percutáneo de la válvula aórtica por acceso transcava

#### To the Editor,

An 86-year-old man, with previous coronary artery disease (recanalization of chronic total occlusion of the right coronary artery) and left ventricular ejection fraction of 45%, presented with symptomatic severe aortic stenosis. Surgical risk was considered high (Society of Thoracic Surgery score 8.1%) and a decision was made to perform transcatheter aortic valve replacement (TAVI).

Preprocedural multislice computed tomography (MSCT) showed diffuse and severe calcified peripheral artery disease. The distal abdominal aorta showed mural thrombosis and calcification, with an effective diameter of 8.0 mm. Transfemoral access was prevented by diffuse heavily calcified bilateral iliac and femoral artery stenosis, with minimum lumen diameter of 4.5 mm on the right and 4.3 mm on the left. Axillary artery access was unsuitable due to significant peripheral artery disease. Due to previous thoracic radiation and chronic obstructive pulmonary disease, the transapical and direct aortic approaches were not considered as optimal alternatives to the femoral route for this patient and so the possibility of transcaval approach was evaluated. Preprocedural MSCT confirmed eligibility for transcaval TAVI, ruling out anatomical constraints and limiting conditions.<sup>1</sup>

The technique of transcaval access and closure, which has been described previously,<sup>2–4</sup> and retrograde transcatheter aortic valve replacement using a standard transfemoral technique and MSCT-fluoroscopy fusion imaging guidance are shown in [figure 1](#). After simultaneous caval and aortic angiograms ([video 1 of the supplementary data](#)), a 3D segmented model with associated

landmarks (green circles marked the planned transverse puncture site, yellow circles marked vertebrae) was registered on fluoroscopy: anteroposterior ([figure 1A](#)) and lateral ([figure 1B](#)) fluoroscopic views confirming correct position of the catheter in the vena cava and snare in the aorta. An electrified guidewire crossed the aorta wall at the planned level (green circles, [figure 1C](#)), and was trapped in the aortic lumen by the snare ([figure 1D](#), [videos 2 and 3 of the supplementary data](#)). CoreValve Evolut PRO 29 mm was implanted (yellow circles marked the native aortic annulus plane, [figure 1E](#)). To close the transcaval access, an Amplatzer duct occluder device was deployed using a deflectable catheter to achieve perpendicular deployment to the aorta during pullback ([figure 1F](#), [video 4 of the supplementary data](#)). The immediate aortic angiogram after closure showed minimal aortocaval flow ([figure 1G](#)). The final aortogram showed no residual aortocaval flow or contrast extravasation ([video 5 of the supplementary data](#)). Postprocedural MSCT showed an aortocaval fistula and ruled out other vascular complications of transcaval access ([figure 1H](#)). Follow-up MSCT after 30 days demonstrated complete closure of the fistula ([figure 1I](#)).

To the best of our knowledge, the use of MSCT-fluoroscopy fusion imaging (HeartNavigator system; Philips Healthcare, The Netherlands) to facilitate transcaval TAVI has not been previously reported. MSCT is crucial for assessment of eligibility for transcaval TAVI and to determine the optimal caval and aortic puncture site. During transcaval TAVI, MSCT-fluoroscopy fusion imaging determines the optimal fluoroscopy angulations, leads to a reduction in fluoroscopy time and volume contrast and improves procedure safety, since it provides a 3D “road map” for the intervention.

At the present time, MSCT is the gold standard imaging technique for TAVI procedural planning. MSCT-fluoroscopy fusion imaging provides an alternative tool for procedural planning and guidance.