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EDITORIAL

Managing Patients With Advanced Atrioventricular Block: The Essential Role of Cardiovascular Magnetic Resonance Imaging for Timely and Accurate Diagnosis

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Unlike conduction disease in elderly patients, advanced atrioventricular block in young and middle-aged patients is predominantly attributable to causes other than degenerative conduction disease and coronary artery disease.[1] Frequent causes of advanced atrioventricular block in young people include cardiac sarcoidosis,[2–4] giant cell myocarditis,[2] genetic cardiomyopathies[5] caused by mutations in *LMNA*,[6] *SCN5A*,[7] and *EMD*,[8] and Lyme carditis in places where Lyme disease is endemic.[9]

See Article by Vuorinen et al.

Identifying the cause of advanced atrioventricular block is critical because it guides clinical management, the cause-specific treatments are often distinct, which in turn influences long-term outcomes.[10] Unfortunately, the cause of advanced atrioventricular block in young people is often unrecognized, particularly in the absence of extracardiac manifestations of a systemic disease, such as sarcoidosis. In a large Danish study of 1027 patients aged <50 years who received their first permanent pacemaker between 1996 and 2015, the cause of the advanced atrioventricular block was unknown in 50%.[11] The consequences of not knowing the cause of the advanced atrioventricular block were described in a follow-up study: these patients had a 3- to 4-fold increase in the risk of the composite end point of death, hospitalization for heart failure, ventricular tachyarrhythmia, or aborted cardiac arrest at a median follow-up of 9.8 years when compared with a cohort of matched controls without pacemakers.[12]

The underlying cause of advanced atrioventricular block can frequently be identified with the help of cardiovascular magnetic resonance imaging (CMR)[13,14]; in the study by Baritussio et al, CMR provided incremental diagnostic value in 65% of young and middle-aged patients with advanced atrioventricular block.[13] The presence of subepicardial, septal-predominant, multifocal late gadolinium enhancement on CMR would indicate cardiac sarcoidosis[15] as the underlying cause, whereas the absence of late gadolinium enhancement would definitively exclude cardiac sarcoidosis and giant cell myocarditis, allowing the clinician to look for causes of advanced atrioventricular block that could present without late gadolinium enhancement, such as Lyme carditis and degenerative conduction disease (Figure).

Key Words: Editorials ■ atrioventricular block ■ magnetic resonance imaging

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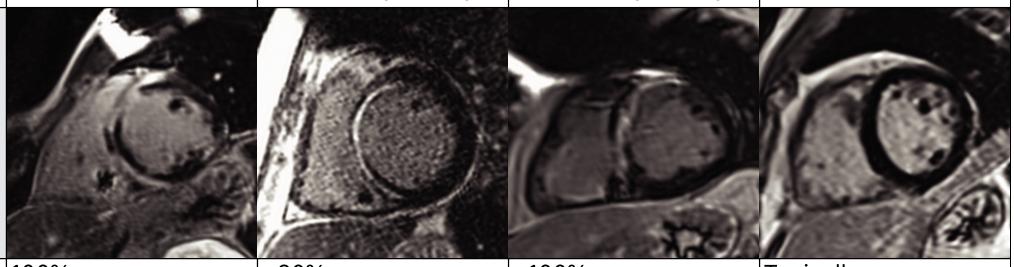
	Cardiac sarcoidosis	*LMNA* cardiomyopathy	Ischemic cardiomyopathy	Lyme carditis
Example LGE images				
LGE prevalence	100%	~60%	~100%	Typically none
Typical LGE pattern	Subepicardial or transmural, multifocal, septal-predominant	Midmyocardial, involving the septal and the inferior segments	Subendocardial or transmural, involving a coronary artery territory	None
Typical medical treatment	Immunosuppression and GDMT for HF/systolic dysfunction	GDMT for HF/systolic dysfunction	GDMT for HF/systolic dysfunction and the secondary prevention of CAD	Antibiotics
Typical device treatment	ICD	ICD	Permanent pacemaker	No permanent device

Figure. Example cardiovascular magnetic resonance imaging of patients with advanced atrioventricular block from 4 causes with distinct treatments.
CAD indicates coronary artery disease; GDMT, guideline-directed medical therapy; HF, heart failure; ICD, implantable cardioverter-defibrillator; and LGE, late gadolinium enhancement.

The main challenge in doing CMR to diagnose the cause of advanced atrioventricular block is that patients may need continuous pacing while they get evaluated, and conventional temporary transvenous pacemakers may not be safe for magnetic resonance imaging. In this context, Vuorinen et al describe, in this issue of the *Journal of the American Heart Association* (*JAHA*), the use of temporary permanent pacemakers (TPPMs) to allow CMR in patients with advanced atrioventricular block requiring temporary pacing.[16] TPPM, which involves the use of a temporary active-fixation lead with an externalized permanent generator taped to the skin, was originally devised as a solution for temporary pacing in pacemaker-dependent patients requiring total removal of infected devices[17] and is an established method of temporary pacing in patients with a contraindication for immediate permanent pacemaker implantation.[18] Vuorinen et al show that TPPMs could allow CMR to be performed safely and effectively. No patient in the study had CMR-related adverse events, and the TPPM generator did not lead to any significant artifacts on the CMR images. Among 17 patients, CMR led to a diagnosis of histologically confirmed granulomatous inflammatory cardiomyopathy in 8, including cardiac sarcoidosis in 7 and giant cell myocarditis in 1. Another patient was suspected of having cardiac sarcoidosis based on CMR, but histological confirmation could not be obtained.

Over the past several years, a wealth of data have demonstrated the safety of magnetic resonance imaging in patients with cardiac implantable electronic devices, including those considered to be non–magnetic resonance conditional.[19] Because TPPM involves the use of leads and generators that are designed to be used permanently, the patients can be scanned with the same safety precautions as those with permanent pacemakers. Although susceptibility artifacts are a consideration when doing CMR on patients with devices, they are less of a concern with permanent pacemakers compared with implantable cardioverters-defibrillators; with right-sided pacemakers, all conventional CMR sequences can be performed without any impact on image quality.[20] As described by Vuorinen et al, TPPMs on the right side should not result in any cardiac artifacts related to the generator. Thus, TPPMs should preferentially be placed on the right side to allow high-quality CMR, including in patients in whom permanent devices would be implanted on the left side.

Although their experience was limited to only 17 patients, Vuorinen et al have indubitably demonstrated that CMR with TPPM is feasible, is safe, and can help diagnose cardiac pathologies with important clinical implications. Timely and accurate diagnosis of the underlying cause of advanced atrioventricular block by the routine use of CMR, particularly in young and middle-aged patients, will enable cause-specific treatments, and improve outcomes. Using TPPM in patients who need continuous pacing will make this clinical practice easier.

ARTICLE INFORMATION

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