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Management of high-degree atrioventricular block in the setting of cardiac sarcoidosis: a therapeutic controversy

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To the Editor,

Cardiac sarcoidosis (CS) is generally considered as an underdiagnosed condition that might emerge in isolation or as part of a systemic disease involving lungs, skin, liver, etc. (1-4). Importantly, this phenomenon might be complicated by heart failure, malignant tachyarrhythmias as well as variable degrees of atrioventricular block (AVB) (1-4). In their recently published article (1), Obi ON, et al. have focused on a variety of controversial topics in the management of CS. Accordingly, we would like to make a few comments on the management of high-degree AVBs in patients with CS.

In the setting of CS, clinicians are generally inclined to implant cardiac devices (permanent pacemakers or implantable cardioverter-defibrillators (ICDs)) in the early stage of the disease course once the general indications for these invasive therapeutic modalities are apparently met. Accordingly, high-degree AVBs are immediately managed with permanent pacemaker implantation (PPCI) in most cases with active CS. However, there might exist a potential likelihood of AVB recovery (complete or partial) in response to antiinflammatory regimens in certain patients with CS (4-9). In this context, previous reports (5-9) demonstrated successful management of second and third degree AVBs with steroid therapy in the setting of CS without further need for PPCI. Of note, steroid therapy might be highly efficacious for AVB management particularly when it is started in the early stages of CS (1, 5-8). In general, it may be suggested that 'the shorter the duration of AVB, the higher the chance of steroid response may be likely' in the setting of CS (8). Moreover, initiation of high-dose intravenous steroid pulse for a few days (methylpredonisolone 1g/day) followed by oral steroids (rather than oral steroids alone) might serve as a more efficient strategy for AVB management in patients with active CS (8). However, the issue of whether combination therapy (steroid plus methotrexate, etc.) or specific antiinflammatory agents (infliximab, etc). (1) might have a more favorable impact on the recovery of AVB still remains to be fully established. On the other hand, temporary cardiac pacing might be necessary as a bridge to AVB recovery (9) or PPCI in hemodynamically unstable patients while they are under antiinflammatory medication.

Importantly, it also seems possible to predict therapeutic response of high-degree AVBs on cardiac imaging in patients with CS (4,5,7). Certain findings including myocardial inflammation (as might be evidenced by fluorodeoxyglucose (FDG) uptake) around the atrioventricular (AV) nodal structures might potentially suggest AVB recovery in response to steroid therapy (4,5,7). In this context, it seems reasonable to carry out decision-making for PPCI after a short period of steroid therapy. However, late gadolinium enhancement (LGE) near these structures without signs of inflammation (on magnetic resonance imaging (MRI)) might immediately warrant PPCI in those with high-degree AVBs on admission largely attributable to the poor response to steroid therapy in this setting (4,5).

Notably, Heart Rhythm Society (HRS) guideline highly encourages ICD implantation in patients

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with CS requiring PPCI (Class IIa) due to the potential risk of malignant arrhythmias in these patients (2,3). However, pacemaker devices have been preferred by some clinicians in CS patients without an overt indication for ICD therapy (5) possibly due to the higher complication rates of ICD devices (inappropriate shocks, etc.). Finally, intra or infra-Hisian AVBs might have the potential to recur after successful management with steroid therapy in patients with CS, and requires close follow-up (4). In this regard, this might potentially warrant advanced tests including electrophysiological study (EPS) in an attempt to carry out decision-making for prophylactic PPCI (potentially indicated in those with residual infra-Hisian abnormalities presenting with a prolonged H-V interval on EPS) (4).

In summary, potential over-treatment with PPCI is quite likely in patients with active CS presenting with high-degree AVBs. In this context, early initiation of antiinflammatory agents (in higher initial doses and possibly in combination) might significantly reduce the need for PPCI in select cases with CS (4-9). This might obviate device-related potential complications particularly in the young population (2). However, clinical follow-up seems to be mandatory in CS patients with a high-degree AVB succesfully managed with antiinflammatory agents, particularly with regard to AVB recurrence in the long term (4).

Conflicts of Interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

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