Slow pathway modification for treatment of pseudo-pacemaker syndrome due to first-degree atrioventricular block with dual atrioventricular nodal physiology

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Introduction
Marked first-degree atrioventricular (AV) block related to dual AV nodal physiology can result in atrial contraction coinciding with ventricular systole and “pseudo-pacemaker syndrome.” To date, literature regarding the treatment of this rare condition has been limited to dual-chamber pacing to restore a physiologic AV interval. A potential concern for performing slow pathway modification in these patients is uncertainty regarding residual fast pathway conduction.

Case report
A 58-year-old man without evidence of structural heart disease presented for evaluation of palpitations and “neck throbbing” with mild exertion. Physical examination (while asymptomatic) was unrevealing. Electrocardiography demonstrated normal sinus rhythm with first-degree AV block (PR 245 ms) and left anterior fascicular block (QRS 105 ms) (Figure 1A). Ambulatory rhythm monitoring revealed symptoms associated with episodes of abrupt PR prolongation > 400 ms with higher sinus rates, which was replicated in the office (Figure 1B). The patient was scheduled for pacemaker implantation at another institution and sought a second opinion. Risks and benefits of both slow pathway modification and pacemaker implant were discussed, including the possibility of requiring pacemaker implantation following slow pathway modification.

Figure 1 Marked abrupt first-degree atrioventricular (AV) block occurring with ambulation is suggestive of dual AV nodal physiology. Baseline electrocardiograms: A: at rest; B: with mild exertion.

KEYWORDS Pacemaker syndrome; Pseudo-pacemaker syndrome; Symptomatic first-degree AV block; Slow pathway; Ablation

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KEY TEACHING POINTS

- Marked first-degree atrioventricular (AV) block is a well-recognized cause of the pseudo-pacemaker syndrome, whereby atrial systole in close proximity to the preceding ventricular systole leads to various hemodynamic changes, including systemic hypotension, elevated pulmonary arterial pressures, and cannon A-waves. In instances of dual AV nodal physiology, preferential slow pathway conduction can present in a similar fashion.

- Literature regarding the treatment of this rare condition has been limited to dual-chamber pacing to restore a typical AV mechanical relationship.

- A potential concern for performing slow pathway modification in patients with pseudo-pacemaker syndrome due to preferential slow pathway conduction is inadequate residual fast pathway conduction.

- Electrotonic coupling between fast and slow pathway has been demonstrated, with complete ablation of the slow pathway associated with shortening of fast pathway effective refractory period.

- We describe a case of pseudo-pacemaker syndrome due to preferential slow pathway conduction that was successfully treated with slow pathway ablation. This was associated with improvements in fast pathway Wenckebach cycle length and PR interval (at rest and with exertion). The latter finding remains robust and the patient remains asymptomatic after 9 months of follow-up.

Subsequent invasive electrophysiological study under light sedation demonstrated AH 110 ms, HV 52 ms, AV Wenckebach cycle length 590 ms, fast pathway effective refractory period (ERP) of 610 ms, and slow pathway ERPs of 470 ms and 430 ms while pacing at a drive cycle length of 850 ms. With incremental pacing the PR interval abruptly prolonged from 240 ms at a paced cycle length (PCL) of 820 ms to 365 ms at a PCL of 800 ms (Figure 2A). With low-dose isoproterenol, the PR interval approached the R-R interval at physiological PCLs (Figure 2B). There was no sustained tachycardia inducible with atrial or ventricular programmed stimulation with isoproterenol infusion up to 20 µG/min. Slow pathway modification was performed with application of radiofrequency energy (Supplemental Figure 1) during isoproterenol infusion to facilitate antegrade fast pathway conduction. Subsequently, following isoproterenol washout, 1:1 fast pathway conduction was present at PCL as short as 730 ms, with a PR interval of 200 ms. This finding remained robust with isoproterenol infusion (Figure 2C). There was no longer evidence of dual AV nodal physiology. Subsequent ambulatory rhythm monitoring revealed heart rates as high as 98 beats/min with 1:1 AV conduction and stable PR interval 220–230 ms with complete resolution of symptoms (Figure 3A and B). He remains asymptomatic without further intervention after 9 months of subsequent follow-up.

Discussion

First-degree AV block is typically an asymptomatic electrocardiographic finding; however, its presence has been associated with elevated rates of atrial fibrillation, the need for permanent pacemakers, and mortality, with the burden of risk proportional to the degree of PR interval prolongation. Marked first-degree AV block is a rare but well-recognized cause of the “pseudo-pacemaker syndrome,” whereby atrial systole in close proximity to the preceding ventricular systole leads to various hemodynamic changes, including systemic hypotension, elevated pulmonary arterial pressures, and cannon A-waves. Dual AV nodal physiology with preferential antegrade slow pathway conduction can present in a similar fashion. Inadvertent fast pathway injury during catheter ablation for AV nodal reentrant tachycardia has also been associated with the pseudo-pacemaker syndrome.

To date, the treatment of this condition has been dual-chamber pacemaker implantation to restore a physiologic AV interval. In case reports and small uncontrolled trials including patients with similar symptoms and with PR intervals >300 ms, this intervention is associated with symptomatic and functional improvement and is recommended in the current guidelines for device-based therapies of cardiac rhythm abnormalities (IIa, level of evidence B).

Given that preferential slow pathway conduction can result in a markedly prolonged PR interval and the pseudo-pacemaker syndrome, slow pathway modification may represent a therapeutic alternative to permanent pacing in selected patients. It is possible that this approach would leave the patient with insufficient residual fast pathway conduction, thus necessitating pacemaker placement; however, there are data to suggest that elimination of electrotonic inhibition of the fast pathway by slow pathway modification will facilitate fast pathway conduction. Strickberger and colleagues demonstrated that complete loss of slow pathway function after slow pathway modification for AV nodal reentrant tachycardia was associated with a significant improvement in fast pathway conduction, whereas fast pathway conduction was unchanged after slow pathway modification with residual slow pathway function. Consistent with these data, we found that following elimination of slow pathway conduction we
observed a reduction in fast pathway ERP and PR shortening with 1:1 fast pathway conduction at substantially shorter cycle length than prior to ablation. The shortening of PR interval (both at rest and with exertion) appears robust in the 9 months following the patient’s procedure and he remains asymptomatic to date.

Conclusion
To our knowledge, this is the first report of slow pathway modification to treat symptomatic first-degree AV block occurring in the setting of dual AV nodal physiology. That this treatment can improve macroscopic electrophysiological properties of the fast pathway, a finding that appears robust after extended follow-up, suggests it may represent a viable therapeutic alternative to permanent pacing.

Appendix
Supplementary data
Supplementary data associated with this article can be found in the online version at https://doi.org/10.1016/j.hrcr.2017.10.003.
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