

# A case of premature ventricular contraction originating at the aortomitral fibrous continuity and exiting from the anterolateral papillary muscle due to muscular chordae tendineae



Yoichiro Nakagawa, MD, Hiroshi Furusho, MD, PhD, Kenji Miwa, MD, PhD, Toshihiko Yasuda, MD, PhD

*From the Department of Cardiology, Ishikawa Prefectural Central Hospital, Ishikawa Prefecture, Japan.*

## Introduction

Most idiopathic premature ventricular contractions (PVCs) originate from the right ventricular outflow tract, and catheter ablation is an established treatment option.<sup>1</sup> In contrast, idiopathic PVCs originating from the left ventricle (LV) can be difficult to treat with catheter ablation. Idiopathic PVCs originating from the LV are frequently associated with conduction systems, such as the Purkinje fiber network, and have been reported to form a unique and complex arrhythmic substrate.<sup>2</sup>

Catheter ablation of PVCs originating from the left ventricular papillary muscle (LVPM) was first reported by Dopalapudi and colleagues in 2008.<sup>3</sup> Owing to the subsequent development of electrocardiogram diagnostic prediction algorithms and imaging technologies such as 3D mapping, diagnostic accuracy and treatment outcomes for PVCs originating from the LVPM have gradually improved. However, the true mechanism of this arrhythmia is still largely unknown. A recent report suggested that papillary muscles can serve as preferential conduction via Purkinje fibers inside,<sup>4</sup> and the origin can be remote from the exit.

An isolated prepotential could be an indicator of successful catheter ablation in patients with idiopathic PVC originating from the aortomitral fibrous continuity (AMFC).<sup>5</sup> However, AMFC is usually composed of anatomically strong fibrous tissue connecting the aortic annulus and the mitral annulus. Therefore, given the lack of myocardial fibers, it is unclear why AMFC can be the origin of PVC.<sup>6</sup> Although AMFC has a fibrous structure that connects with the anterior mitral leaflet (AML), its boundaries are inconspicuous and

## KEY TEACHING POINTS

- Idiopathic premature ventricular contractions (PVCs) originating from the left ventricle are frequently associated with the Purkinje network and form a complex arrhythmic substrate. In particular, the true mechanisms underlying PVCs originating from the papillary muscles and aortomitral fibrous continuity (AMFC) are still unknown.
- Abnormalities in the myocardial delamination process may result in the formation of chordae tendineae containing the embryological remnants of myocardial fibers. Such muscular chordae tendineae may be involved in the conduction system and arrhythmic substrate of the left ventricular papillary muscle group.
- Given the lack of myocardial fibers, it is still unclear why AMFC can be the origin of PVC, but the embryological remnants of myocardial tissue in the AMFC region may serve as an arrhythmic substrate.

difficult to distinguish via computed tomography or echocardiography. This differentiation is difficult even in catheter ablation; thus, we collectively refer to this area as  $AML_{AMFC}$ .

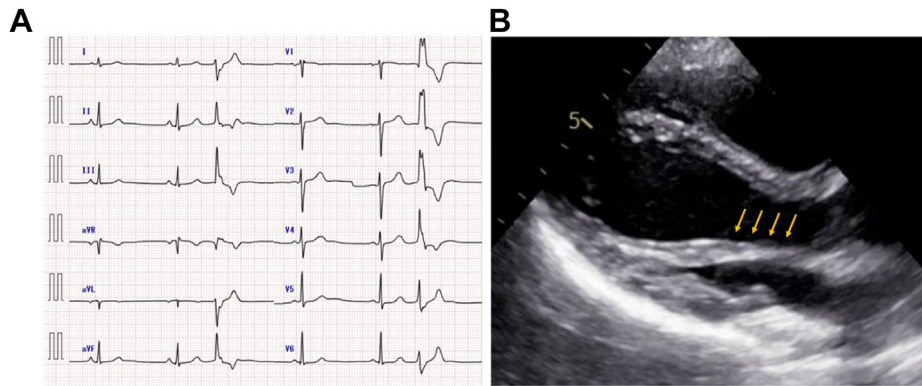
Despite the proximity, there are few reports discussing the mechanism and the treatment of idiopathic PVCs associated with LVPM, chordae tendineae, and  $AML_{AMFC}$ . Here, we report a rare case of idiopathic PVC originating from the  $AML_{AMFC}$  and exiting from the anterolateral papillary muscle (ALPM).

## Case report

A 48-year-old female patient without any past medical history was referred to our hospital for frequent PVCs. The patient had no history of smoking or excessive alcohol or

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Funding Sources: None. Disclosures: The authors have no conflicts of interest to declare. **Address reprint requests and correspondence:** Dr Yoichiro Nakagawa, Department of Cardiology, Ishikawa Prefectural Central Hospital, 2-1 Kuratsuki Higashi, Kanazawa City, Ishikawa Prefecture, Japan 920-8530. E-mail address: [yoichiro.n.99@gmail.com](mailto:yoichiro.n.99@gmail.com).



**Figure 1** Electrocardiogram and echocardiographic findings. **A:** Electrocardiogram showing premature ventricular contraction with right bundle branch block pattern and right inferior axis. **B:** Echocardiography showing a long-axis image of the left ventricle in the diastolic phase. Relatively thick anterolateral papillary muscle chordae tendineae attached to anterior mitral leaflet were observed.

caffeine intake. Physical examination showed no signs of heart failure. Blood tests showed a slightly elevated brain natriuretic peptide level of 57.1 pg/mL, but no other abnormalities were found. The PVC was monomorphic, and the morphology was right bundle branch block and right inferior axis. Based on the previous report,<sup>7</sup> ALPM was suspected as the origin (Figure 1A). Holter electrocardiogram showed 29,332 beats of monomorphic PVC per day. This is equivalent to 27.0% of the total heartbeats. It occurred more frequently from night to morning, and >3 lasting beats were not confirmed. Echocardiography revealed a 60.4% ejection fraction, no enlargement of chamber size or valvular disease, and relatively thick ALPM chordae tendineae attached to the AML (Figure 1B). Given the lack of structural heart disease, this PVC was diagnosed as idiopathic. Because of the failure in suppressing the PVCs and the worsening of symptoms by oral beta blocker, we performed a catheter ablation.

### Catheter ablation

Frequent PVCs were observed throughout the case. The CARTO® 3 system (Biosense Webster, Inc, Irvine, CA) was used for 3D mapping, and the right internal jugular vein, right femoral veins, and right femoral artery were punctured under local anesthesia. A decapolar catheter (EPStar; Japan Lifeline Co, Ltd, Tokyo, Japan) was inserted into the coronary sinus, and another decapolar catheter (Scooper; FUKUDA DENSHI Co, Ltd, Tokyo, Japan) was placed into the right ventricle to record both His and right ventricle electrogram.

An intracardiac ultrasound catheter (SOUNDSTAR; Biosense Webster, Inc) was used to create an anatomical shell of the LV, the aortic valve, and the papillary muscles at the beginning of the procedure. A trabeculated structure was noted from the ALPM to the AML (Figure 2A). Then, a mapping catheter (PentaRay; Biosense Webster, Inc) was inserted into the LV for the activation mapping of the PVC. Contrary to what we anticipated, the earliest activation was at the AML<sub>AMFC</sub> (Figure 2B), where a discrete prepotential preceded the QRS onset of the PVC by 84 ms (Figure 2C). This site

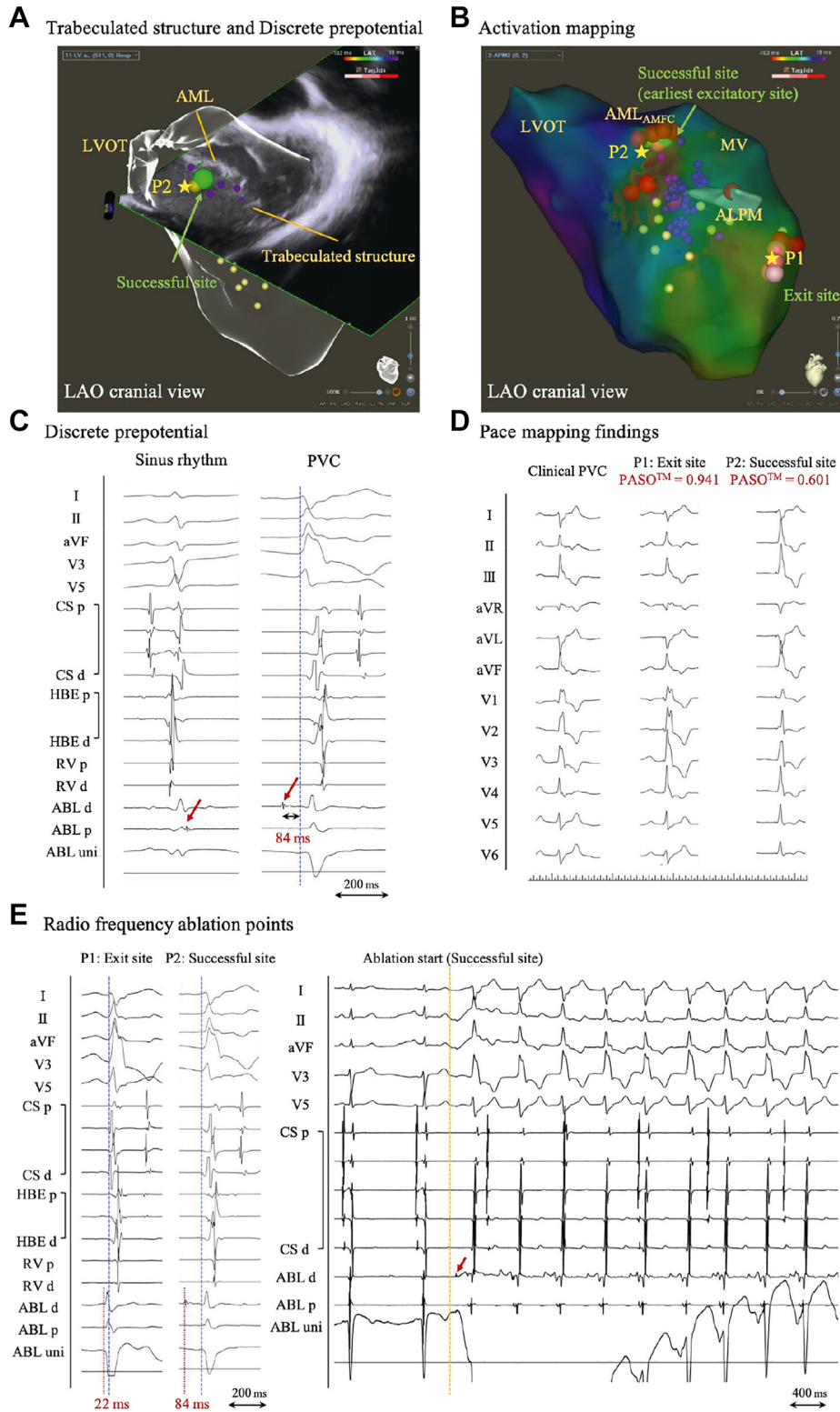
coincided with the trabeculated structure noted by the intracardiac ultrasound (Figure 2A). The discrete potential noted during PVC could be observed after the offset of the QRS during sinus rhythm, which suggested the retrograde conduction via the trabeculated structure (Figure 2C). No prepotential was noted around the ALPM, and the 3D mapping showed later activation (Supplemental Video 1).

Pace mapping score obtained by the PaSo™ module (Biosense Webster, Inc) was the best at the base of the ALPM; however, the score was low at the AML<sub>AMFC</sub> (Figure 2D). Radiofrequency (RF) ablation was first applied using a 3.5 mm irrigation catheter (THERMOCOOL SMART-TOUCH™ SF; Biosense Webster, Inc) at the ALPM where the best pace mapping score was obtained (Figure 2E). However, this failed to suppress the PVCs.

Considering the instability of the ablation catheter and the possible intramural origin, RF ablation was performed at the tip of the ALPM. However, this also failed to suppress the PVCs. Given the evidence of the discrete prepotential at the AML<sub>AMFC</sub> and the result of pace mapping, we postulated that the origin of this PVC was at the AML<sub>AMFC</sub> and the exit was at the ALPM. RF application at the AML<sub>AMFC</sub> successfully eliminated the PVC after accelerated PVCs with the same morphology (Figure 2E). No PVC was observed after RF ablation even with isoproterenol infusion.

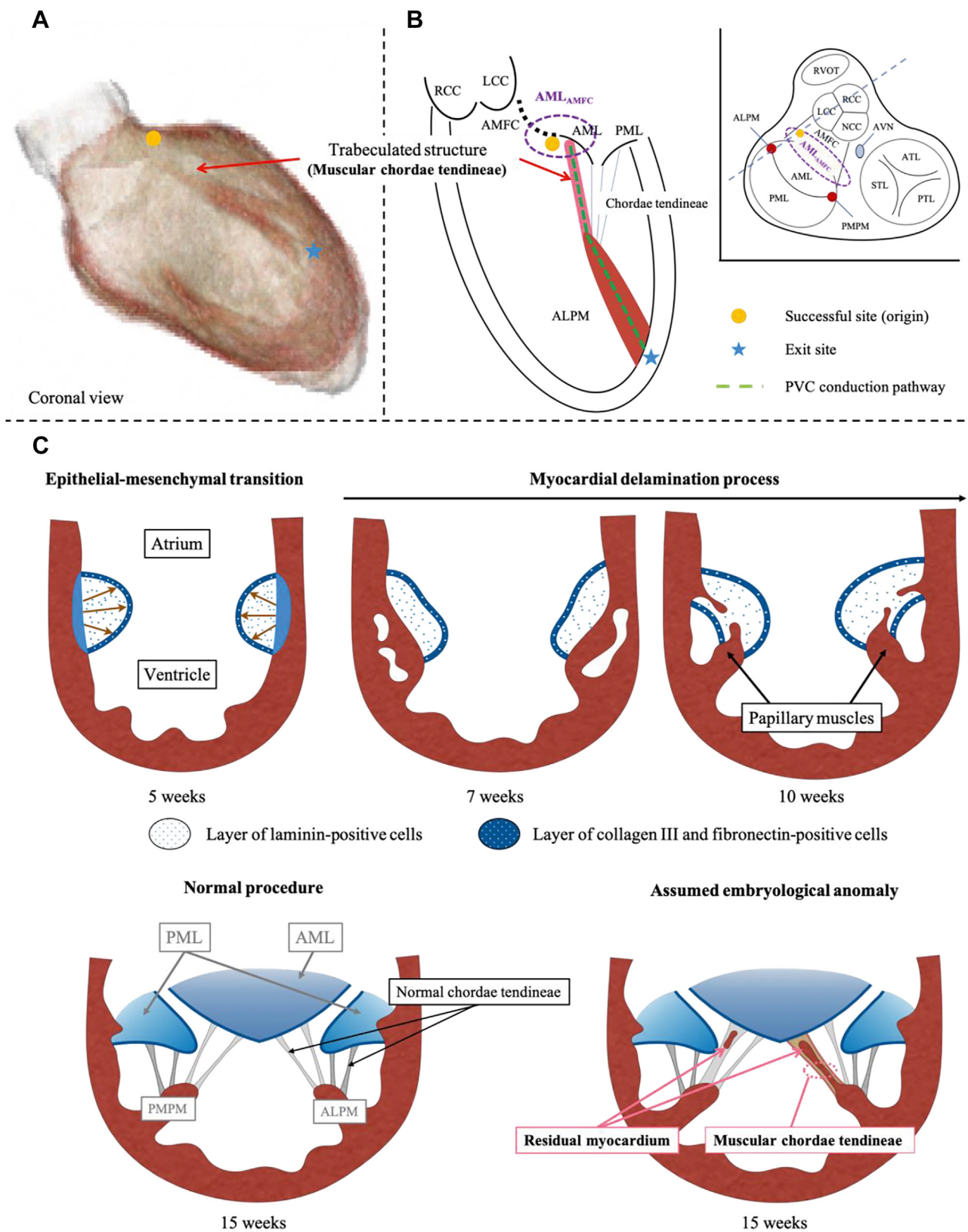
### Discussion

In this present case, the PVC originated at the AML<sub>AMFC</sub> and exited from the ALPM. RF application targeting the discrete potential successfully eliminated the PVC even though the pace mapping did not match. This suggested the existence of a preferential pathway connecting the AML<sub>AMFC</sub> and the ALPM. A reconstructed enhanced computed tomography image also showed evidence of a remarkable trabeculated structure connecting the AML<sub>AMFC</sub> and ALPM (Figure 3A). The presumed preferential pathway connecting the origin and exit of the PVC is depicted in Figure 3B. The trabeculated structure must contain remnants of myocardial tissue, given the evidence of the discrete prepotential along the structure. Papillary muscles, chordae tendineae, and



**Figure 2** Findings during catheter ablation. **A:** A trabeculated structure that continuously binds to the anterior mitral leaflet (AML) was confirmed by intracardiac ultrasound. Along this structure, discrete prepotential was recorded from the aortomitral fibrous continuity (AMFC) to the anterolateral papillary muscle (ALPM). **B:** The activation map of the premature ventricular contraction (PVC) exhibited the earliest activation site at the AML<sub>AMFC</sub> region. The exit site of the PVC was on the basal side of the ALPM. **C:** The discrete prepotential recorded at the distal electrodes of ablation catheter leading to PVC was observed up to 84 ms earlier than the onset of QRS. This potential was observed as a delayed potential behind the QRS during sinus rhythm. **D:** The pace mapping score was the highest on the ALPM basal side, but very low in the AMFC region. **E:** PVC was not eliminated and QRS morphology did not change during radiofrequency ablation at the ALPM, which was considered the exit site. When RF ablation was applied to the AML<sub>AMFC</sub>, accelerated PVC was observed. The QRS morphology of the accelerated PVC was identical to that of clinical PVC. ABL = ablation catheter; CS = coronary sinus HBE = His bundle electrogram; LVOT = left ventricular outflow tract; MV = mitral valve; PASO<sup>TM</sup> = pace mapping software (CARTO3; Biosense Webster, Inc, Irvine, CA), RV = right ventricle. Yellow tag: pace mapping points; purple tag: prepotential points; red or pink tag: radiofrequency application points.





atrioventricular valve are formed approximately 5–15 weeks after embryogenesis via epithelial-mesenchymal transition and myocardial delamination,<sup>8</sup> as shown in Figure 3C. The leaflets and chordae tendineae are formed from the cell layer

of collagen III and fibronectin-positive cells and the basal layer of laminin-positive cells developed via epithelial-mesenchymal transition. The myocardium that remains in this area during delamination gradually retracts toward the

annulus and papillary muscle, eventually disappearing and becoming fibrous tissue. The myocardial tissue remaining in the chordae tendineae can form an arrhythmic substrate and participate in excitatory transmission as a result of the abnormal delamination process.

The involvement of the “dead-end tract,” which is the embryological remnant of the conduction system, has been suggested as one of the underlying mechanisms.<sup>9</sup> In addition to this hypothesis, we believe that the embryological remnant of myocardial tissue in the AMFC region may serve as an arrhythmic substrate. The prepotential preceding PVC was recorded along the trabeculated structure. The discrete prepotential recorded at AML<sub>AMFC</sub> during PVC was recognized after the offset of QRS during sinus rhythm, which was considered as the retrograde conduction through Purkinje fibers inside the preferential pathway. A previous study has shown that the papillary muscle is anatomically rich in Purkinje fibers and forms various reentry circuits.<sup>10</sup> Although various theories for the development of the conduction system have been proposed, peripheral Purkinje fibers develop differently from the upper conduction system. Purkinje fibers are modified by signals from surrounding tissues, and the local myocardium is differentiated and formed.<sup>11</sup>

In a previous study, Lam and colleagues<sup>12</sup> reported chordae tendineae containing whole or locally myocardial tissue as “muscular chordae tendineae.” There is also a report of a thick muscular mitral chord obstructing the left ventricular outflow tract.<sup>13</sup> Although these reports support our hypothesis, the phenomenon was most likely caused by the muscular chordae tendineae in the present case. According to another study, such chordae tendineae are found in approximately 14.65% of the population.<sup>14</sup> Most false tendons are composed of fibrous tissue, but 5.3% of them are of the muscular type, including myocardial tissue and Purkinje fibers.<sup>15</sup> The false tendon does not attach to the mitral valve leaflets, but the muscular chordae tendineae in the present case are thought to have properties similar to the muscular type of false tendon. As far as we know, this is the first report indicating that the residual myocardium in the chordae tendineae served as the preferential pathway of PVC. Although the electrical connection between AML<sub>AMFC</sub> and ALPM was poorly understood, we believe that this is a significant finding to help understand ventricular arrhythmias around this area.

## Conclusion

We report a rare case of PVC originating from the AML<sub>AMFC</sub> and exiting from the basal side of the ALPM. The embryolog-

ical remnants of myocardial fibers in chordae tendineae were suspected as the underlying mechanism.

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## Appendix Supplementary Data

Supplementary data associated with this article can be found in the online version at <https://doi.org/10.1016/j.hrcr.2022.09.017>.

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**Figure 3** Cardiac computed tomography image, schematic views of the premature ventricular contraction (PVC) conduction pathway, and peripheral structure and development of the papillary muscle. **A:** Reconstructed cardiac computed tomography images confirmed myocardial bundle tissue connected to the AML<sub>AMFC</sub>. **B:** PVC originated at the AML<sub>AMFC</sub> region and was assumed to exit from the left ventricle via the muscular chordae tendineae and ALPM. **C:** After epithelial-mesenchymal transition, papillary muscles, chordae tendineae, and atrioventricular valve are formed via myocardial delamination approximately 5–15 weeks after embryogenesis. The leaflets and chordae tendineae are formed from a basal layer of laminin-positive cells and a cell layer of collagen III and fibronectin-positive cells. Myocardium remaining in this area gradually retracted toward the annulus and papillary muscles and finally disappeared. In this case, it is suggested that residual myocardium remained in the chordae tendineae. It has been reported that the residual myocardium can be anatomically observed as a “muscular chordae tendineae,” which is much thicker than any other chordae. ALPM = anterolateral papillary muscle; AMFC = aortomitral fibrous continuity; AML = anterior mitral leaflet; ATL = anterior tricuspid leaflet; AVN = atrioventricular node; LCC = left coronary cusp; NCC = noncoronary cusp; PML = posterior mitral leaflet; PMPM = posteromedial papillary muscle; PTL = posterior tricuspid leaflet; RCC = right coronary cusp; RVOT: right ventricular outflow tract; STL = septal tricuspid leaflet.