

Images in  
Cardiovascular Medicine



# A Case of SCN5A Mutation-Associated Isolated Left Atrial Standstill and Ischemic Stroke

Dong-Seon Kang , MD\*, Pichmanil Khmao , MD\*, Jaewon Oh , MD, PhD, and Hui-Nam Pak , MD, PhD

Division of Cardiology, Department of Internal Medicine, Yonsei University College of Medicine, Yonsei University Health System, Seoul, Korea



**Received:** Mar 24, 2022  
**Revised:** May 12, 2022  
**Accepted:** Jun 8, 2022  
**Published online:** Jun 24, 2022

### Correspondence to

Hui-Nam Pak, MD, PhD

Division of Cardiology, Department of Internal Medicine, Yonsei University College of Medicine, Yonsei University Health System, 50-1, Yonsei-ro, Seodaemun-gu, Seoul 03722, Korea.





Email: hnpak@yuhs.ac

\*Dong-Seon Kang and Pichmanil Khmao contributed equally to this work and share first authorship.

Copyright © 2022. The Korean Society of Cardiology

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<https://creativecommons.org/licenses/by-nc/4.0>) which permits unrestricted noncommercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

### ORCID iDs

Dong-Seon Kang   
<https://orcid.org/0000-0001-9468-7517>  
Pichmanil Khmao   
<https://orcid.org/0000-0001-6376-6108>  
Jaewon Oh   
<https://orcid.org/0000-0002-4585-1488>  
Hui-Nam Pak   
<https://orcid.org/0000-0002-3256-3620>

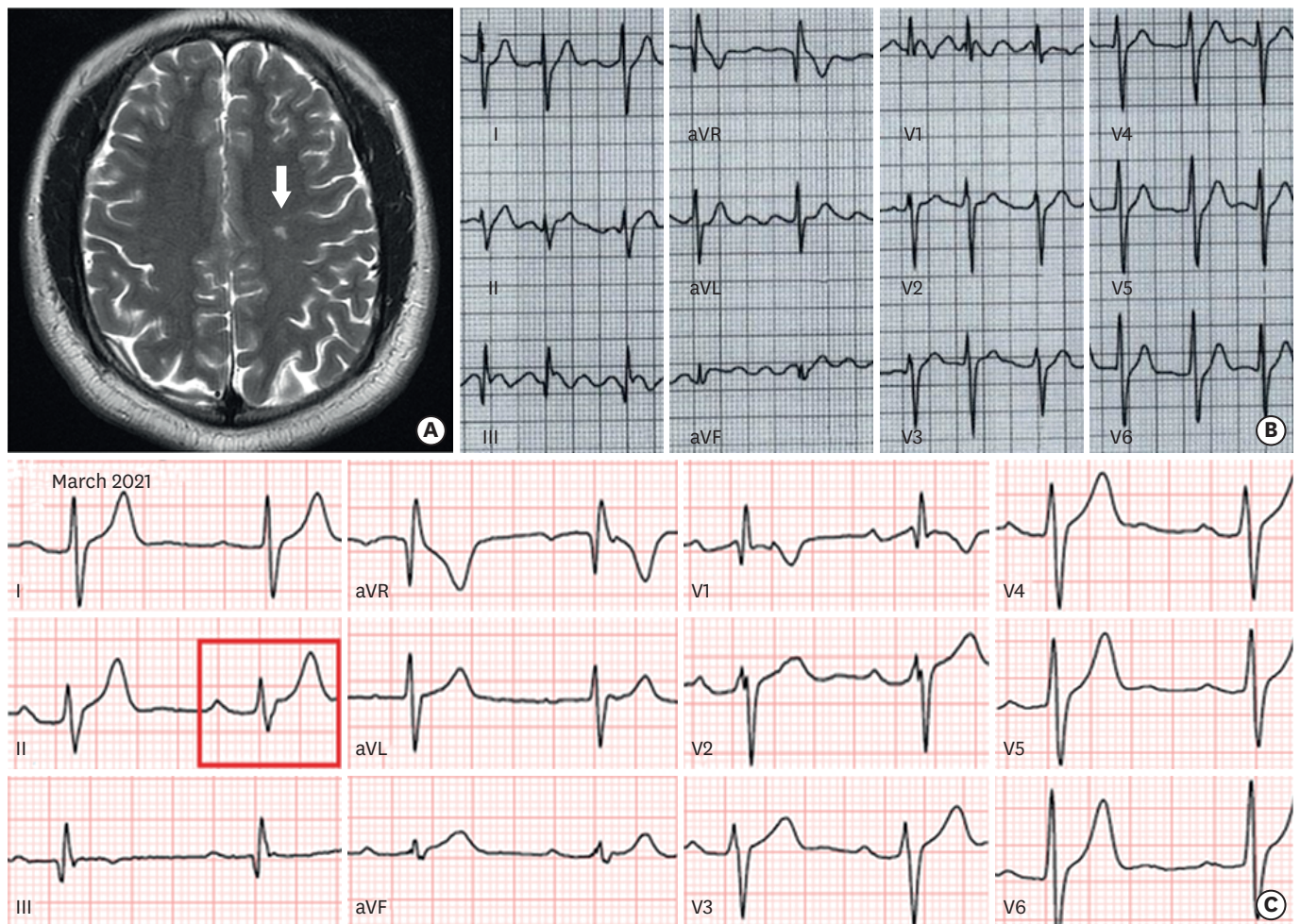
A 26-year-old man experienced paroxysmal atrial fibrillation (AF) and cerebral infarction without significant head and neck angiopathy or coagulopathy in April 2020 (**Figure 1A and B**). The patient has a history of recurrent syncope and was referred to manage a flecainide-resistant paroxysmal AF in July 2020. Electrocardiography and echocardiography revealed PR prolongation (**Figure 1C**) and progressive disappearance of mitral inflow A-wave (**Figure 2**), respectively. Other echocardiography parameters are shown in **Supplementary Figure 1A and B**.

The patient underwent an electrophysiological study in March 2021. At a baseline sinus cycle length of 700 ms, the HV interval was 73 ms (**Figure 3A**). LA pressure was measured at 33 mmHg. Intracardiac echocardiography was performed after trans-septal puncture (**Supplementary Figure 1C**). In contrast to the absence of endocardial bipolar voltage signals in the entire LA and pulmonary veins except for a small area at the anteroseptal LA (a bipolar voltage cut-off of 0.1mV, **Figure 3B**), right atrium (RA) showed relatively preserved voltage with localized scar on the septum in a 3D-electroanatomical map. The intracardiac electrograms showed consistent results (**Figure 3A**). We performed linear ablations of the superior vena cava to the RA septum and cavotricuspid isthmus without LA ablation.

Two months after the procedure, an *SCN5A* mutation (c.3823G>A[p.Asp1275Asn], likely pathogenic) was found in the proband and his mother and sister (**Figure 3C**). Paroxysmal complete atrioventricular block was revealed in the Holter during follow-up (**Figure 3D**). Eventually, the patient underwent a dual-chamber pacemaker implantation eight months after electrophysiology procedure.

Our patient had an *SCN5A*-D1275N mutation associated with isolated LA standstill and progressive AV conduction disease requiring permanent pacemaker implantation.<sup>1)2)</sup> Although this patient did not show any evidence for ventricular arrhythmia, careful observation and follow-up are warranted for this potentially progressive genetic arrhythmia disease.

Written informed consent was obtained from the patient.



**Figure 1.** Examinations which were conducted prior to electrophysiological study. Cerebral magnetic resonance imaging in April 2020 (A). Documented paroxysmal atrial fibrillation in July 2020 (B). Electrocardiogram in March 2021. The red square showed PR interval 240 ms (C). aVF = augmented vector foot; aVL = augmented vector left; aVR = augmented vector right.

**Funding**

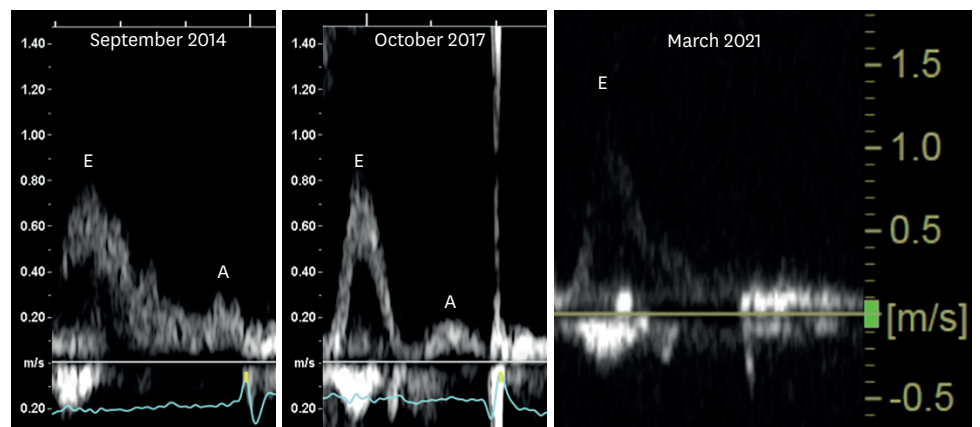
This work was supported by a grant (HI21CO011) from the Ministry of Health and Welfare, a grant (NRF-2020R1A2B5B01001695) from the Basic Science Research Program run by the National Research Foundation of Korea (NRF) which is funded by the Ministry of Science, ICT & Future Planning (MSIP), and a grant (RS-2022-00141473) from Cross-ministerial tasks.

**Conflict of Interest**

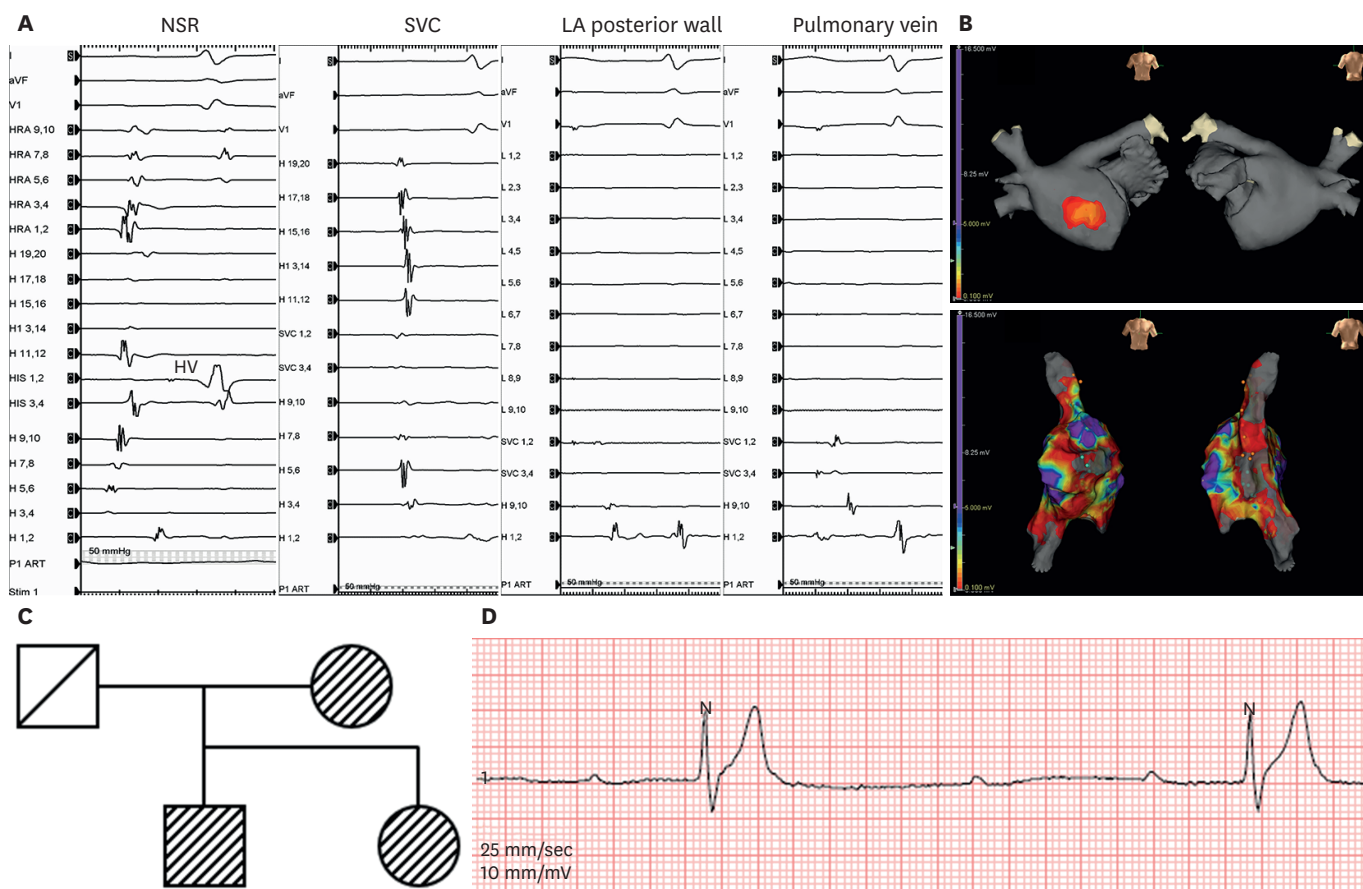
The authors have no financial conflicts of interest.

**Data Sharing Statement**

The data generated in this study is available from the corresponding author upon reasonable request.



**Figure 2.** Echocardiography which was conducted in March 2021. Serial change of mitral inflow pattern and loss of A-wave since September 2014.



**Figure 3.** Electrophysiologic study and ECG. Intracardiac electrogram during sinus rhythm with HV interval 73 ms. In contrast to the absence of endocardial signals in LA posterior wall and pulmonary vein, SVC showed relatively preserved signals in intracardiac electrograms (A). The left atrial voltage map with final ablation site at RA (B). A genotype pedigree representing first-degree relatives (C). Follow-up Holter showing paroxysmal complete AV block in November 2021 (D). AV = atrioventricular; aVF = augmented vector foot; HV = His-ventricular; LA = left atrial; NSR = normal sinus rhythm; RA = right atrium; SVC = superior vena cava.

**Author Contributions**

Conceptualization: Oh J, Pak HN; Data curation: Kang DS, Khmao P, Oh J, Pak HN; Formal analysis: Kang DS, Khmao P, Pak HN; Investigation: Kang DS, Khmao P, Oh J, Pak HN; Methodology: Kang DS, Khmao P, Pak HN; Project administration: Pak HN; Supervision: Oh J, Pak HN; Writing - original draft: Kang DS, Khmao P; Writing - review & editing: Oh J, Pak HN.

**SUPPLEMENTARY MATERIAL**

**Supplementary Figure 1**

Additional results of examinations which was conducted prior to or during electrophysiological study. Transthoracic echocardiography conducted in March 2021 identified an enlarged left ventricle with preserved systolic function (A). Other parameters of the transthoracic echocardiography (B). Intracardiac echocardiography showed decreased emptying and filling velocity of left atrial appendage (C).

[Click here to view](#)

**REFERENCES**

1. Shaw TR, Northridge DB, Francis CM. Left atrial standstill in a patient with mitral stenosis and sinus rhythm: a risk of thrombus hidden by left and right atrial electrical dissociation. *Heart* 2003;89:1173. [PUBMED](#) | [CROSSREF](#)
2. Duncan E, Schilling RJ, Earley M. Isolated left atrial standstill identified during catheter ablation. *Pacing Clin Electrophysiol* 2013;36:e120-4. [PUBMED](#) | [CROSSREF](#)