




Cerebral Infarction from Sick Sinus Syndrome (Based on *SCN5A*) via Patent Foramen Ovale

Shuang He^{1,2,3,4,5} · Lei Zhang^{1,2,3,4,5} · Xiao-Yan Liu^{1,2,3,4,5} · Xiao-Juan Ji^{2,3,4,5,6} · Shi-Long Tang^{2,3,4,5,7} · Xiao-Yun Wu^{1,2,3,4,5} · Tie-Wei Lu^{1,2,3,4,5} 

Received: 21 March 2023 / Accepted: 20 April 2023 / Published online: 5 May 2023
© The Author(s), under exclusive licence to Dr. K C Chaudhuri Foundation 2023

To the Editor: A 9-y-old girl with hemiplegia as the initial symptom, suddenly developed left limb paralysis without obvious inducement and was admitted to our hospital. Cerebral infarction, characterized by significant reduction in the lumen of branches of the right middle cerebral artery, was confirmed through Cranial MRI and MRA. Meanwhile, the patient had recurrent bradycardia (sinus bradycardia/arrest) – tachycardia (atrial tachycardia/flutter) by detected by ECG and Holter monitoring and was positive for atropine test. Transesophageal ultrasound showed patent foramen ovale (PFO), and contrast echocardiography of the right heart indicated right-to-left shunt. Additionally, genetic examination suggested that the patient's *SCN5A* gene had a heterozygous mutation (c.719dupT), leading to change in the corresponding amino acids (p. A242Gfs*11).

Based on her clinical manifestations, imaging and genetic examination, the patient was diagnosed as sick sinus syndrome (SSS). A thrombus formed in the right atrium due to abnormal heart rhythm entered the systemic circulation

through the small PFO, thereby resulting in cerebral artery embolism and, eventually, hemiplegia of the left limb. To avoid severe damage recurrence, the patient received radiofrequency ablation to cure tricuspid isthmus-dependent atrial flutter, interventional occlusion of the foramen ovale, double chamber pacemaker implantation to correct sinus bradycardia/arrest, after above treatment, and her condition obviously improved through 2 y regular follow-up.

This case is the first report of cerebral infarction caused by SSS in children. SSS caused by ion channel-related gene abnormalities was the pathophysiological basis of cerebral infarction in this case [1, 2]. PFO is a potential pathway capable of causing cerebral infarction; which suggests that relevant examinations should be performed in children with abnormal heart rhythm or nervous system manifestations. Occlusion of the foramen ovale should be recommended as an early intervention to avoid severe complications [3, 4].

Declarations

Conflict of Interest None.

References

1. Alkorashy M, Al-Ghamdi B, Tulbah S, et al. A novel homozygous *SCN5A* variant detected in sick sinus syndrome. *Pacing Clin Electrophysiol.* 2021;44:380–4.
2. John RM, Kumar S. Sinus node and atrial arrhythmias. *Circulation.* 2016;133:1892–900.
3. Mojadidi MK, Zaman MO, Elgendy IY, et al. Cryptogenic stroke and patent foramen ovale. *J Am Coll Cardiol.* 2018;71:1035–43.
4. Regmi MR, Lara Garcia OE, Parajuli P, Madrid O, Hafiz AM. Late atrial thrombus formation after percutaneous patent foramen ovale closure. *JACC Case Rep.* 2020;2:636–40.

Publisher's Note Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

✉ Tie-Wei Lu
ltw200145@163.com

- 1 Department of Cardiovascular Medicine, Children's Hospital of Chongqing Medical University, 136 Second Zhongshan Road, Chongqing 400014, China
- 2 Ministry of Education Key Laboratory of Child Development and Disorders, Chongqing, China
- 3 National Clinical Research Center for Child Health and Disorders, Chongqing, China
- 4 China International Science and Technology Cooperation Base of Child Development and Critical Disorders, Chongqing, China
- 5 Chongqing Key Laboratory of Pediatrics, Chongqing, China
- 6 Department of Ultrasound, Children's Hospital of Chongqing Medical University, Chongqing, China
- 7 Department of Radiology, Children's Hospital of Chongqing Medical University, Chongqing, China