

Intraatrial reentrant tachycardia originating from the prior suture line of the baffle in a patient who underwent the Mustard operation: Ultra-high-density 3-dimensional mapping



Jae-Sun Uhm, MD, PhD, Hee Tae Yu, MD, PhD, Tae-Hoon Kim, MD, Boyoung Joung, MD, PhD, Hui-Nam Pak, MD, PhD, Moon-Hyoung Lee, MD, PhD

From the Division of Cardiology, Department of Internal Medicine, Severance Hospital, Yonsei University College of Medicine, Seoul, Republic of Korea.

Introduction

The incidence of arrhythmia is higher in patients with congenital heart disease than in those with a structurally normal heart.¹ Scar-related arrhythmias are common long after cardiac surgery for congenital heart disease.² The Mustard operation is a kind of an atrial switch operation that was performed in patients with transposition of the great arteries (TGA). However, the incidence of atrial tachyarrhythmia is high in patients who have undergone this operation.³ We describe a case of intraatrial reentrant tachycardia (IART) originating from the prior suture line of the baffle in a patient who had undergone the Mustard operation 10 years previously. To treat the IART, ultra-high-density 3-dimensional (3-D) electroanatomic mapping and radiofrequency catheter ablation (RFCA) were performed.

Case report

A 32-year-old man visited the outpatient clinic because of recurrent palpitations and dizziness. At birth, he was diagnosed as having TGA. He underwent pulmonary arterial banding at the age of 2 years and the Mustard operation (with the use of autologous pericardial tissue) at the age of 22 years. He underwent percutaneous intervention for stenosis of the superior baffle at the age of 24 years. Electrocardiography performed at the outpatient clinic revealed atrial flutter and a ventricular rate of 92 beats/min (Figure 1). The function and size of both ventricles were found to be normal on echocardiography. His atrial flutter and symptoms persisted despite medical therapy. We decided to perform

an electrophysiological study and RFCA by using an ultra-high-density 3-D electroanatomic mapping system (Rhythmia; Boston Scientific, Marlborough, MA) and an Orion catheter (Boston Scientific) to treat the atrial flutter. Cardiac computed tomography was performed before the electrophysiological study (Supplemental Figure 1). In the electrophysiological laboratory, the atrial flutter was sustained. A decapolar catheter (Woven catheter; Boston Scientific) and a His-RV catheter (Japan Lifeline, Tokyo, Japan) were respectively placed into the coronary sinus and morphologic left ventricle through the inferior baffle. The tachycardia cycle length was 260 ms. Systemic venous atriography was performed. Under the guidance of fluoroscopy and intracardiac echocardiography, the inferior baffle was punctured with a manually shaped Brockenbrough needle and an SL-1 introducer, according to previously described procedures.⁴ After the baffle puncture, pulmonary venous atriography was performed. Next, ultra-high-density 3-D mapping of the systemic venous atrium was performed during tachycardia. During activation mapping, double potentials were observed on the inferior baffle wall, near the inferior vena cava, which corresponded to the prior suture line between the baffle and the atrium (Figure 2). Ultra-high-density 3-D activation and propagation maps revealed figure-of-8 IART involving the inferior baffle (Figure 3A, Supplemental Video, and Supplemental Video Snapshot). The critical isthmus of the reentrant circuit with slow conduction was located on the inferior baffle near the inferior vena cava, which corresponded to the area of the double potentials. Another small reentry circuit was observed at the area about 2 cm superior to the previously described isthmus. 3-D activation mapping of the pulmonary venous atrium could not be completed because the tachycardia degenerated into atrial fibrillation during 3-D mapping at the right pulmonary veins. Linear ablation was performed 2 times along the isthmus of the reentry circuit and other small reentry circuit by using an irrigated ablation catheter (Figure 3). After RFCA, we confirmed that there were no electrical signals along the

KEYWORDS Arrhythmia; Atrial tachycardia; Congenital heart disease; Intraatrial reentrant tachycardia; Mustard operation; Rhythmia system; Transposition of the great arteries; Ultra-high-density mapping (Heart Rhythm Case Reports 2018;4:451–454)

Address reprint requests and correspondence: Dr Moon-Hyoung Lee, Division of Cardiology, Severance Cardiovascular Hospital, Yonsei University College of Medicine, 50-1 Yonsei-ro Seodaemun-gu, Seoul, 03722 Korea. E-mail address: mhlee@yuhs.ac.

KEY TEACHING POINTS

- The incidence of atrial tachyarrhythmia is high in patients who have undergone the Mustard operation.
- It is highly probable that patients who underwent this operation have an extensive scar along the prior long suture lines in the atria.
- The prior suture lines of the baffle can be an arrhythmogenic substrate in these patients.
- An ultra-high-density 3-dimensional mapping system is useful for visualization of the reentry circuit of intraatrial reentrant tachycardia in these patients.

ablation line. RFCA of the cavotricuspid isthmus (CTI) was performed in both the systemic and pulmonary venous atria. Bidirectional block of the CTI was confirmed by pacing at the coronary sinus and lower portion of the lateral wall of the systemic venous atrium (Supplemental Figure 2). Tachycardia was not induced by incremental and ramp pacing. Tachycardia did not recur in the 8 months after RFCA.

Discussion

TGA is characterized by an abnormal arrangement of the aorta and the main pulmonary artery and accounts for 5% to 7% of all congenital heart diseases.⁵ The Mustard operation was developed for atrial switch with intraatrial baffles in patients with TGA in the 1960s.⁶ However, the operation has been known to have sequelae, including atrial tachyarrhythmia, sinus node dysfunction, progressive systemic

ventricular failure, and sudden cardiac death.^{3,7} In the 1980s, the Mustard operation was replaced by the arterial switch operation. Most of the patients who had undergone the Mustard operation have become adults. Among them, a considerable number of patients have experienced the sequelae of the operation. In the Mustard operation, pericardial tissue was sutured with the atria for the formation of the baffles from both caval veins to the morphologic left ventricle.⁸ Therefore, long suture lines in the atria were inevitable. In the long term, the suture lines become fibrotic and have the potential to be an arrhythmogenic substrate.^{9,10}

For the management of atrial tachyarrhythmia in patients who underwent the Mustard operation, physicians need to pay attention to the risk of sinus node dysfunction, systemic ventricular dysfunction, and sudden cardiac death. Because the sinus node and its blood supply could be damaged during the operation,⁷ the prevalence of sick sinus syndrome is high.⁹ Symptoms of sick sinus syndrome can appear after termination of tachycardia, especially during administration of antiarrhythmic drugs. Patients who underwent the Mustard operation can be susceptible to hemodynamic instability or systemic ventricular dysfunction because their morphologic right ventricle works as the systemic ventricle.⁷ Furthermore, patients with atrial tachyarrhythmia and systemic ventricular dysfunction are at a high risk of sudden cardiac death.^{11,12} Use of antiarrhythmic drugs is limited, because antiarrhythmic drugs have suppressive effects on the sinus node activity and negative inotropic effects. Therefore, RFCA plays a major role in the treatment of tachycardia in these patients.

RFCA is a challenging procedure in patients who underwent the Mustard operation because of 2 hurdles: the complex cardiac anatomy (and the potential necessity for baffle puncture) and extensive scar in the atria in these patients. Preprocedural cardiac imaging studies including

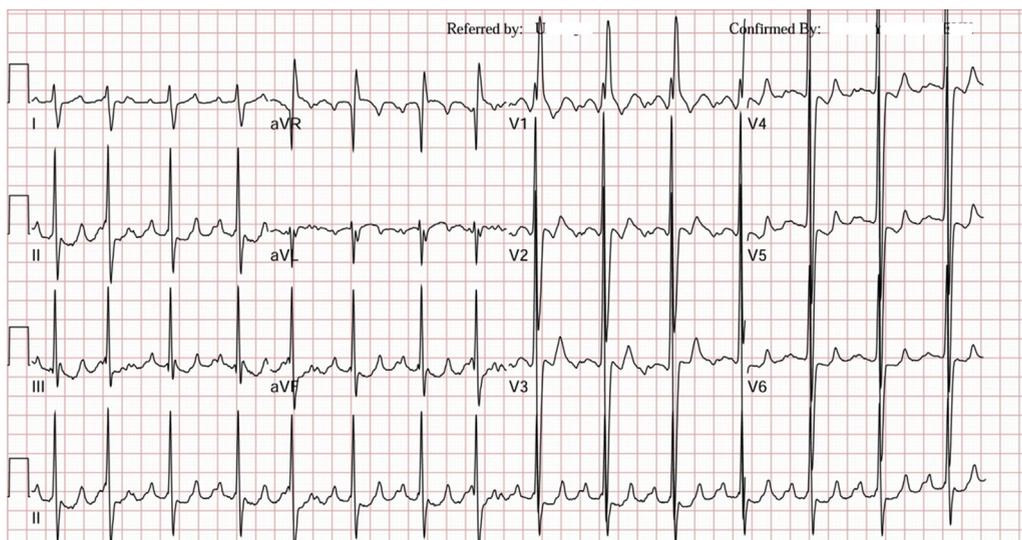


Figure 1 Electrocardiogram obtained in the outpatient clinic.



Figure 2 Intracardiac electrogram obtained during the tachycardia. Double potentials were recorded at spline B 4-5 of the Orion catheter.

echocardiography, computed tomography, and magnetic resonance imaging are necessary for understanding of the cardiac anatomy. A detailed review of prior operation records and a discussion with the cardiac surgeon are important. The coronary sinus ostium is usually located in the systemic venous atrium for maintenance of physiologic circulation. In some cases, the coronary sinus ostium is located in the pulmonary venous atrium because the surgeon made the baffle beside the coronary sinus ostium to avoid damage to the conduction system. The baffle puncture needs preparation. The baffle puncture techniques have been described in a previous article.⁴ Intracardiac echocardiography is helpful during baffle puncture.

It is highly probable that patients who underwent the Mustard operation have an extensive scar along the prior long suture lines in the atria. Most atrial tachyarrhythmias in these patients are scar-related IART.⁹ It is important to create a detailed 3-D activation and voltage map. The Rhythmia and the Orion catheter enable rapid acquisition and automated annotation of hundreds of electrograms. An ultra-high-density 3-D mapping system is useful for detailed visualization of the reentrant circuit and its critical isthmus. At first, the systemic venous atrium should be mapped rather than the pulmonary venous atrium. This is because the systemic venous atrium is easily accessible and IARTs originating from the systemic venous atrium are more common than those

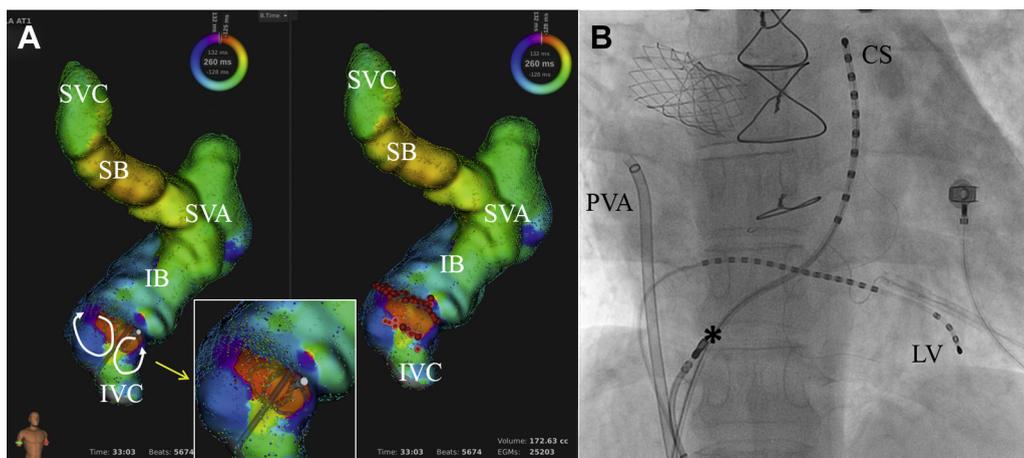


Figure 3 **A:** Ultra-high-density 3-dimensional activation map of the tachycardia showing figure-of-8 intraatrial reentrant tachycardia (white arrows) with the isthmus at the prior suture line at the inferior baffle. Figure in the white box is a magnified image of the reentry circuit. Two ablation lines were formed along the isthmus. The red filled-in circles indicate the ablation lesions. **B:** Fluoroscopic image of the ablation site. The SL-1 introducer was located in the pulmonary venous atrium through the baffle puncture. * indicates radiofrequency catheter ablation site. CS = coronary sinus; IB = inferior baffle; IVC = inferior vena cava; LV = left ventricle; PVA = pulmonary venous atrium; SB = superior baffle; SVA = systemic venous atrium; SVC = superior vena cava.

originating from the pulmonary venous atrium owing to the larger scar area in the systemic venous atrium.¹³ CTI block may be needed even in patients with non-CTI-dependent IART, because the incidence of CTI-dependent atrial flutter is high in these patients.⁹ For a complete CTI block, RFCA should be performed in both the systemic and pulmonary venous atria. This is because the baffle wall is located between the tricuspid annulus and the inferior vena cava.

In the present case, the ultra-high-density 3-D electroanatomic map well visualized the critical isthmus of the reentrant circuit in the inferior baffle. The critical isthmus was compatible with the prior suture line between the native right atrium and the pericardial patch. Although we could not complete the 3-D activation map of the pulmonary venous atrium, the IART was successfully ablated. This was because the reentry circuit involved only the systemic venous atrium.

Conclusion

The prior suture lines of the baffle can be an arrhythmogenic substrate in patients who underwent the Mustard operation. An ultra-high-density 3-D mapping system is useful for visualization of the reentry circuit of IART in these patients.

Appendix

Supplementary data

Supplementary data associated with this article can be found in the online version at <https://doi.org/10.1016/j.hrcr.2018.06.012>.

References

1. Wu MH, Lu CW, Chen HC, Kao FY, Huang SK. Adult Congenital Heart Disease in a Nationwide Population 2000-2014: Epidemiological Trends,

- Arrhythmia, and Standardized Mortality Ratio. *J Am Heart Assoc* 2018; 7:e007907.
2. Labombarda F, Hamilton R, Shohoudi A, et al. Increasing prevalence of atrial fibrillation and permanent atrial arrhythmias in congenital heart disease. *J Am Coll Cardiol* 2017;70:857–865.
3. Baysa SJ, Olen M, Kanter RJ. Arrhythmias Following the mustard and senning operations for dextro-transposition of the great arteries: clinical aspects and catheter ablation. *Card Electrophysiol Clin* 2017;9:255–271.
4. Uhm JS, Kim NK, Kim TH, Joung B, Pak HN, Lee MH. How to perform trans-conduit and transbaffle puncture in patients who have previously undergone the Fontan or Mustard operation. *Heart Rhythm* 2018;15:145–150.
5. van der Linde D, Konings EE, Slager MA, Witsenburg M, Helbing WA, Takkenberg JJ, Roos-Hesselink JW. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. *J Am Coll Cardiol* 2011; 58:2241–2247.
6. Mustard WT. Successful two-stage correction of transposition of the great vessels. *Surgery* 1964;55:469–472.
7. Dos L, Teruel L, Ferreira IJ, Rodriguez-Larrea J, Miro L, Girona J, Albert DC, Goncalves A, Murtra M, Casaldaliga J. Late outcome of Senning and Mustard procedures for correction of transposition of the great arteries. *Heart* 2005; 91:652–656.
8. Konstantinov IE, Alexi-Meskishvili VV, Williams WG, Freedom RM, Van Praagh R. Atrial switch operation: past, present, and future. *Ann Thorac Surg* 2004;77:2250–2258.
9. Khairy P, Van Hare GF. Catheter ablation in transposition of the great arteries with Mustard or Senning baffles. *Heart Rhythm* 2009;6:283–289.
10. Houck CA, Teuwen CP, Bogers AJ, de Groot NM. Atrial tachyarrhythmias after atrial switch operation for transposition of the great arteries: treating old surgery with new catheters. *Heart Rhythm* 2016;13:1731–1738.
11. Kammeraad JA, van Deurzen CH, Sreeram N, Bink-Boelkens MT, Ottenkamp J, Helbing WA, Lam J, Sobotka-Plojhar MA, Daniels O, Balaji S. Predictors of sudden cardiac death after Mustard or Senning repair for transposition of the great arteries. *J Am Coll Cardiol* 2004;44:1095–1102.
12. Khairy P, Harris L, Landzberg MJ, Fernandes SM, Barlow A, Mercier LA, Viswanathan S, Chetaille P, Gordon E, Dore A, Cecchin F. Sudden death and defibrillators in transposition of the great arteries with intra-atrial baffles: a multicenter study. *Circ Arrhythm Electrophysiol* 2008;1:250–257.
13. Kriebel T, Tebbenjohanns J, Janousek J, Windhagen-Mahnert B, Bertram H, Paul T. Intraatrial reentrant tachycardias in patients after atrial switch procedures for d-transposition of the great arteries. Endocardial mapping and radiofrequency catheter ablation primarily targeting protected areas of atrial tissue within the systemic venous atrium. *Z Kardiol* 2002;91:806–817.