JACC: CASE REPORTS VOL. 2, NO. 2, 2020
© 2020 THE AUTHORS. PUBLISHED BY ELSEVIER ON BEHALF OF THE AMERICAN COLLEGE OF CARDIOLOGY FOUNDATION. THIS IS AN OPEN ACCESS ARTICLE UNDER THE CC BY-NC-ND LICENSE (http://creativecommons.org/licenses/by-nc-nd/4.0/).

MINI-FOCUS ISSUE: ELECTROPHYSIOLOGY

INTERMEDIATE

CASE REPORT: CLINICAL CASE

Catheter Ablation of Atrial Tachycardia in a Giant Right Atrium

Thomas Kueffer, MS,[a] Markus Schwerzmann, MD,[b] Alexander Kadner, MD,[b] Laurent Roten, MD[a]

ABSTRACT

A 25-year-old male patient with a giant right atrium presented with atrial tachycardia. Electroanatomic mapping revealed micro–re-entry from a low-voltage zone in the region of the right atrial appendage. Linear ablations across the low-voltage zone terminated the tachycardia. The remaining right atrial tissue was electrically normal. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:230–4) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 25-year-old male patient with a giant right atrium (GRA) that was incidentally diagnosed in childhood presented at our clinic with atrial tachycardia. The tachycardia cycle length was 310 ms with 2:1 atrioventricular conduction (**Figure 1**), resulting in a fixed ventricular heart rate of about 100 beats/min, thus limiting his exercise capacity. The treating physician initiated oral anticoagulation and beta-blocker treatment.

LEARNING OBJECTIVES

- The clinician will understand the role of catheter ablation as an alternative to reduction atrioplasty for tachycardia treatment in a patient with a hugely dilated right atrium.
- The clinician will know the challenges and risk management of an ablation procedure in a hugely dilated right atrium.

INTERVENTIONS

Cardiac imaging, including cardiac magnetic resonance, showed a dilated right atrium measuring 12 cm in the transverse axis and 9.5 cm in the longitudinal axis, mild dilation of the right ventricle, and moderate functional tricuspid valve regurgitation secondary to distortion of the annular plane with malcoaptation of the cusps. No Ebstein's anomaly (i.e., septal apical displacement of the tricuspid valve or other cardiac defects) was present on transthoracic echocardiography and cardiac magnetic resonance (**Figure 2**, Videos 1 and 2) (1). Computed tomography was performed for procedural planning.

MANAGEMENT

Because of the patient's young age, limited exercise capacity, and high probability of arrhythmia recurrence after simple cardioversion, we opted for an invasive rhythm control strategy. However, we were concerned about reports of membrane-like right atrial wall tissue in similar cases. In the heart team, we therefore discussed the following 2 strategies: 1)

From the [a]Department of Cardiology, University Hospital Inselspital, University of Bern, Bern, Switzerland; and the [b]Center for Congenital Heart Disease, University Hospital Inselspital, University of Bern, Bern, Switzerland. The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

Informed consent was obtained for this case.

Manuscript received October 29, 2019; accepted November 5, 2019.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2019.11.039

primary reduction atrioplasty with subsequent catheter ablation in case of recurrence; and 2) direct catheter ablation. We chose the second option because the patient had been asymptomatic so far and because reduction atrioplasty may fail to control the arrhythmia. Additionally, the cavotricuspid isthmus (CTI) may also be part of the arrhythmia and is easily targeted by an ablation procedure.

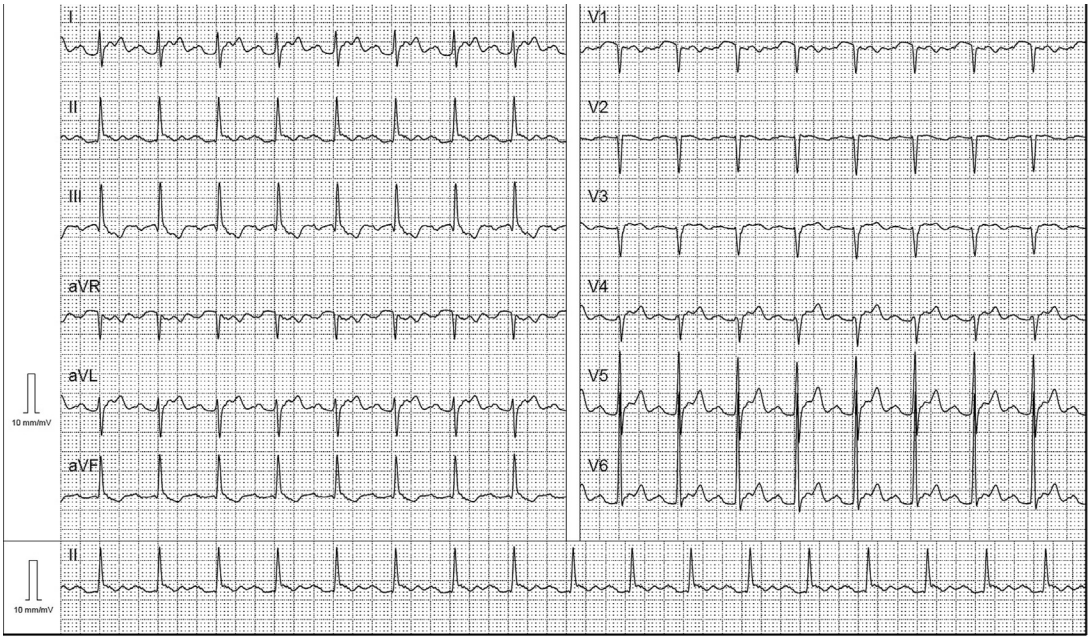
We performed catheter ablation in the electrophysiology laboratory with the patient under general anesthesia and with transesophageal echocardiography during the procedure to monitor the pericardial space. Additionally, we prepared the patient for possible direct sternotomy and femorofemoral cardiopulmonary bypass, as we usually do for lead extraction procedures in the hybrid operating room. Both a cardiac surgeon and a perfusionist with a primed heart-lung machine accompanied the procedure on site. On induction of anesthesia, 1:1 atrioventricular conduction occurred with hemodynamic collapse, necessitating direct current cardioversion. With the patient in sinus rhythm, we reconstructed the right atrium (volume, 638 ml) and the coronary sinus with the Carto 3 System (Biosense Webster, Irvine, California), using a Pentaray eco catheter (Biosense Webster) and a long, steerable sheath (Agilis NxT, large curve, Abbott, Abbott Park, Illinois). Burst stimulation from the proximal coronary sinus very easily induced various atrial tachycardias with changing cycle lengths and different P-wave configurations. Mapping of those tachycardias proved to be impossible because of recurrent, early tactile termination or constantly changing arrhythmias. After lengthy mapping efforts and nonconcurring propagation results, we first ablated a CTI line (length, 7.5 cm) using a Smarttouch SF ablation catheter (Biosense Webster) (Supplemental Figure 1). Eventually, we could show proof of block according to standard criteria (bidirectional block with an interval from the proximal coronary sinus to the CTI line of 180 ms and shorter to the lateral, low right atrium). Afterward, we meticulously acquired a detailed bipolar voltage map, considering only sinus beats. We found a circumscript low-voltage area (<0.5 mV) in the region of the right atrial appendage that was not distinguishable from the right atrium because of the gross dilation (**Figure 3**). In the remaining right atrium, we observed electrically normal tissue. We proceeded with an ablation strategy similar to

ABBREVIATIONS AND ACRONYMS

CTI = cavotricuspid isthmus

GRA = giant right atrium

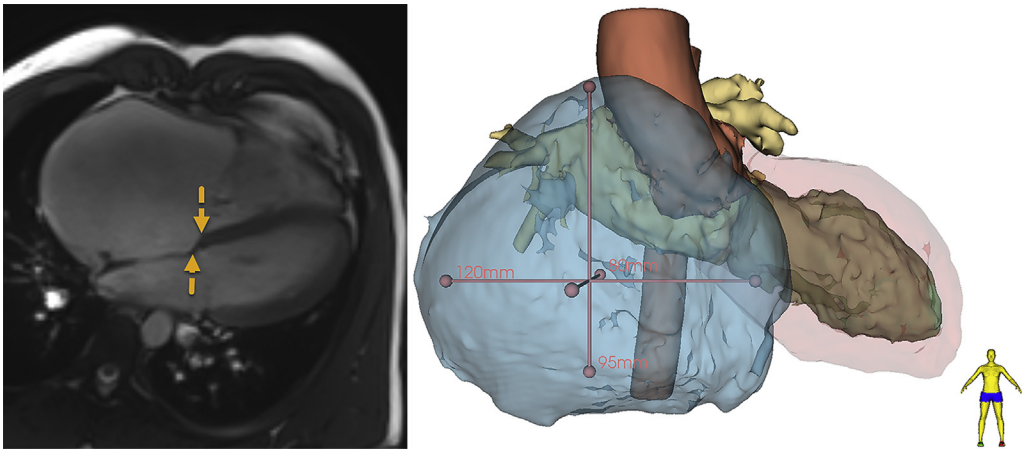
FIGURE 1 12-Lead Electrocardiogram



A 12-lead electrocardiogram of the tachycardia showing atrial flutter with 2:1 atrioventricular conduction and a stable ventricular rate of 100 beats/min. Despite an atypical P-wave configuration, typical flutter cannot be excluded because of the hugely dilated right atrium.

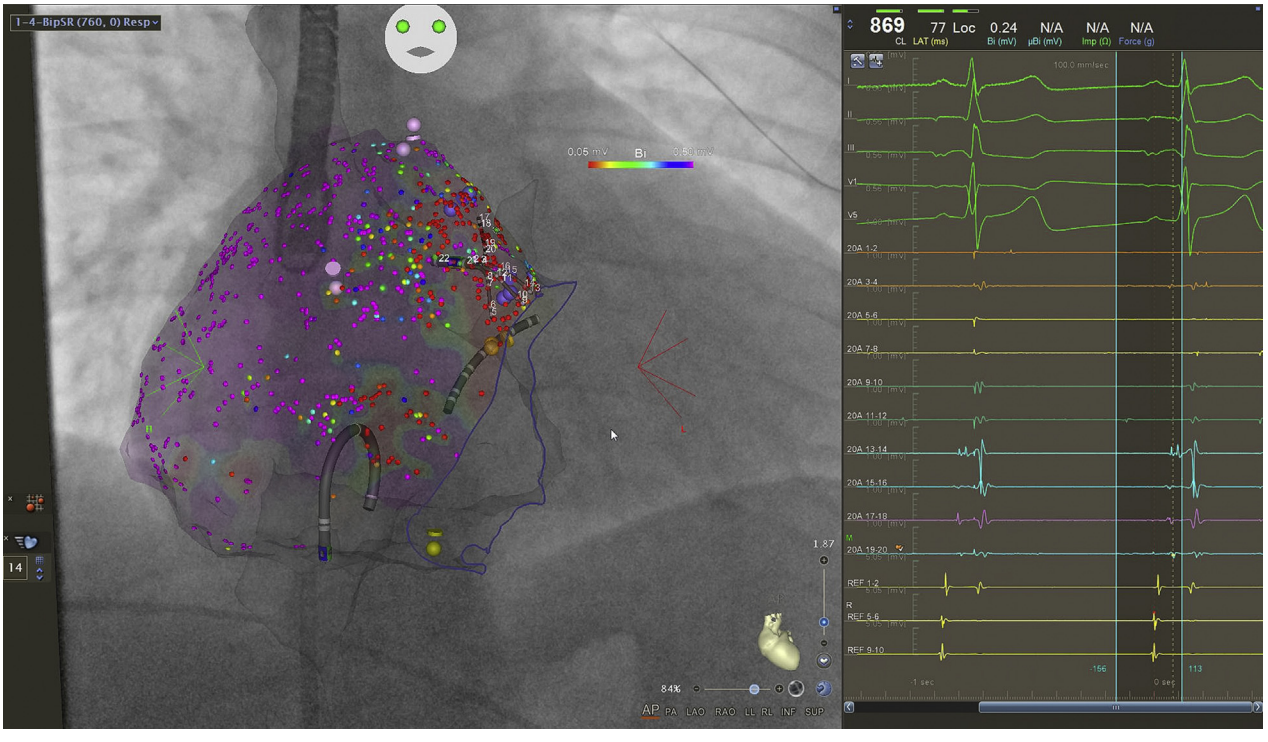
JACC: CASE REPORTS, VOL. 2, NO. 2, 2020
FEBRUARY 2020:230–4

FIGURE 2 Cardiac Imaging



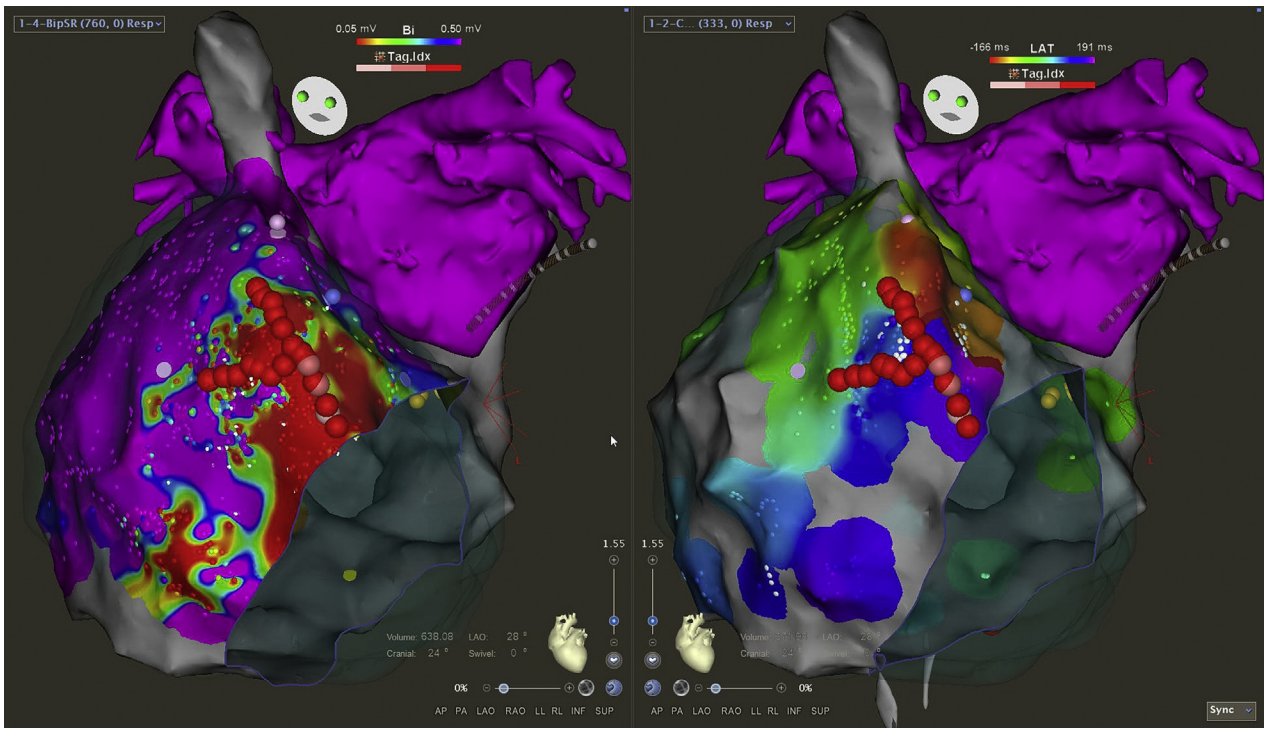
(Left) On cardiac magnetic resonance, no shunt lesion was detected, and the Qp:Qs was 1.0. There was also no major apical displacement **(arrows)** of the septal tricuspid leaflet, as required for the diagnosis of Ebstein's anomaly. **(Right)** On 3-dimensional reconstruction of a computed tomography scan, the enlarged right atrium measured 95 mm in the longitudinal axis, 120 mm in the frontal axis, and 80 mm in the sagittal axis and reached a calculated volume of 611 ml. The 3-dimensional view shows an anteroposterior projection of the right atrium **(blue),** the left ventricular myocardium and aorta **(red),** the left ventricle blood pool, and the left atrium. The image was processed and visualized with Slicer 3D (1). See Videos 1 and 2.

FIGURE 3 Electrograms From the Low-Voltage Area (in Sinus Rhythm)



(Left) Carto (Biosense Webster, Irvine, California) image showing the fluoroscopy image acquired during the procedure with an overlay of the 3-dimensional electroanatomic map acquired in sinus rhythm. The mapping catheter (Pentaray, Biosense Webster) is positioned within the low-voltage zone of the right atrial appendage. **(Right)** Intracardiac electrograms show fractionated and low-amplitude atrial signals.

FIGURE 4 **Voltage and Activation Map**



Carto (Biosense Webster, Irvine, California) images showing **(left)** the voltage map of the giant right atrium, as well as **(right)** the presumed activation map of the tachycardia, processed after ablation. The **red dots** represent the ablation points. The **magenta** left atrium and **transparent blue** right atrium are 3-dimensional segmentations from the computed tomography scan. LAO = left anterior oblique; LAT = local activation time.

substrate modification in ventricular tachycardia ablation. We ablated a line from the tricuspid annulus across the low-voltage area and a second line perpendicular to it. Subsequent aggressive burst stimulation did not induce any further tachycardia. After a waiting period and confirmation of CTI line block, we ended the procedure without complication and without necessitating surgical intervention. After post-processing, the acquired tachycardia maps, we were able to demonstrate micro-re-entry within the low-voltage area as the tachycardia mechanism (**Figure 4**). At 6 months after the procedure, the patient remained asymptomatic, without tachycardia recurrence.

DISCUSSION

Giant right atrium is a rare anomaly with idiopathic, severe dilation of the right atrium. Kurz et al. (2) proposed a right atrial long axis indexed to body surface area >2.6 cm/m^2 for men as a definition. Clearly, our case fulfilled this definition with a value of 5.4 cm/m^2. Other cardiac anomalies, particularly Ebstein's anomaly or tricuspid valve dysplasia, must be excluded for the diagnosis. The most frequent symptoms in patients with GRA are attributed to supraventricular tachycardia and tricuspid regurgitation (3). Many patients remain asymptomatic, but reports of sudden cardiac death in patients with GRA also exist. The underlying pathological process is unclear. In symptomatic cases—mostly because of supraventricular tachycardia—reduction atrioplasty is the predominant treatment strategy and has a high success rate (3–5). No detailed reports on catheter ablation of arrhythmia in GRA have been published. Of note, several surgical reports describe the right atrial wall in such patients to be paper thin and translucent, with absence of myocardium or with lipomatous degeneration (3,5). Catheter manipulation in such patients may lead to right atrial wall perforation or even rupture, particularly because stiff, steerable sheaths are typically used in such dilated cavities for successful catheter manipulation. Interestingly, in our case the right atrial

JACC: CASE REPORTS, VOL. 2, NO. 2, 2020
FEBRUARY 2020:230–4

wall showed only a limited area of low voltage, whereas the remaining right atrium seemed to be electrically normal, a finding suggesting normal right atrial wall tissue. It was not possible to measure right atrial wall thickness in previously acquired cardiac imaging studies with accuracy to determine the procedural risks. Catheter ablation in this case proved to be very challenging because of electrically vulnerable tissue and unstable arrhythmia. We finally succeeded after stepwise ablation of the CTI and additional, substrate-based ablation of the low-voltage area, analogous to ventricular tachycardia ablation. Retrospectively, after post-processing the acquired data, the tachycardia mechanism was compatible with scar-related micro-re-entry.

CONCLUSIONS

Catheter ablation of atrial tachycardia in GRA is challenging but feasible. Attention should be paid to the fact that the right atrial wall may be paper thin in such cases, thus increasing the risk of right atrial wall perforation or even tear. We proposed an approach similar to transvenous lead extraction with conversion to open sternotomy in case of tamponade and subsequent reduction atrioplasty. Alternatively, reduction atrioplasty can be chosen as a first-line strategy, as proposed in the literature.

ADDRESS FOR CORRESPONDENCE: Dr. Laurent Roten, Inselspital, Kardiologie, Freiburgstrasse, 3010 Bern, Switzerland. E-mail: laurent.roten@insel.ch.

REFERENCES

1. Fedorov A, Beichel R, Kalpathy-Cramer J, et al. 3D Slicer as an image computing platform for the quantitative imaging network. Magn Reson Imaging 2012;30:1323–41.

2. Kurz DJ, Oechslin EN, Kobza R, Jenni R. Idiopathic enlargement of the right atrium: 23 year follow up of a familial cluster and their unaffected relatives. Heart 2004;90:1310–4.

3. Binder TM, Rosenhek R, Frank H, Gwechenberger M, Maurer G, Baumgartner H. Congenital malformations of the right atrium and the coronary sinus: an analysis based on 103 cases reported in the literature and two additional cases. Chest 2000;117:1740–8.

4. Forbes K, Kantoch MJ, Divekar A, Ross D, Rebeyka IM. Management of infants with idiopathic dilatation of the right atrium and atrial tachycardia. Pediatr Cardiol 2007;28:289–96.

5. Jonavicius K, Lipnevicius A, Sudikiene R, Zurauskas E, Lebetkevicius V, Tarutis V. Surgical repair of a giant congenital right atrial aneurysm: a case report. J Cardiothorac Surg 2015;10:72.

KEY WORDS 3-dimensional imaging, ablation, congenital heart defect, electroanatomic mapping, idiopathic enlargement, supraventricular arrhythmias

APPENDIX For a supplemental figure and videos, please see the online version of this paper.