

Successful Surgical Treatment of an Electrical Storm in a Patient With Atypical Noncompaction Cardiomyopathy

David Schibilsky, MD; Karin Klingel, MD, MD; Jürgen Schreieck, MD; Meinrad Gawaz, MD; Tobias Walker, MD; Christian Schlensak, MD; Peter Seizer, MD

A 34-year-old male patient with known atypical, predominantly right ventricular noncompaction cardiomyopathy¹ was admitted to our department because of repeated ventricular tachycardias and implantable cardioverter defibrillator shocks (Figure A). An extracorporeal life support system was implanted because of hemodynamic instability, and an electrophysiological study was performed. Epicardial access was achieved by subxiphoid puncture. Endo- and epicardial electro-anatomical voltage mapping (Ensite NavX, St Jude) revealed earliest activation in the area of the noncompaction myocardium (Figure B). Ablation was attempted with radiofrequency (irrigated tip, max 40 Watt) from epi- and endocardial access. However, the patient was still inducible in programmed ventricular stimulation, and during monitoring at the intensive care unit, sustained ventricular arrhythmias occurred. Stepwise, antiarrhythmic treatment including amiodarone, sotalol, and finally ranolazin was used to establish a stable sinus rhythm. However, after >48 hours with stable sinus rhythm, again sustained ventricular tachycardia occurred, and therefore, the patient was allocated to open surgery. Intraoperative epicardial ablation points (see arrows) marked the suspected origin of the arrhythmias (Figure B and C). Accordingly, the arrhythmogenic substrate in the right ventricular apex was completely

removed. Histology and immunohistology showed severe fibrosis in Masson Trichrome stain and infiltration of CD68+ macrophages as arrhythmogenic substrate (Figure D and E). After excision of the noncompaction myocardium, the patient was in stable sinus rhythm. Antiarrhythmic medication was terminated beside amiodarone and β -blocker, and the patient was discharged. Six months after discharge with amiodarone and β -blocker, right ventricular stimulation via the implantable cardioverter defibrillator revealed no ventricular arrhythmia, and thus, antiarrhythmic medication was terminated. The patient was free from heart failure symptoms. Our case emphasizes the importance of an interdisciplinary heart team approach for patients with complex cardiomyopathies.

Disclosures

None.

References

1. Towbin JA, Lorts A, Jefferies JL. Left ventricular non-compaction cardiomyopathy. *Lancet*. 2015;386:813–825. doi: 10.1016/S0140-6736(14)61282-4.

KEY WORDS: cardiomyopathies ■ endocardium ■ heart failure ■ macrophages ■ noncompaction cardiomyopathy

Received January 19, 2016; accepted February 10, 2016.

From the Deutsches Herzkompetenzzentrum, Eberhard Karls-Universität Tübingen, Tübingen, Germany.

Correspondence to Peter Seizer, MD, Medizinische Klinik III, Universitätsklinikum Tübingen, Otfried-Müller Str 10, 72076 Tübingen, Germany. E-mail Peter.Seizer@med.uni-tuebingen.de

(*Circ Heart Fail*. 2016;9:e002973. DOI: 10.1161/CIRCHEARTFAILURE.116.002973.)

© 2016 American Heart Association, Inc.

Circ Heart Fail is available at <http://circheartfailure.ahajournals.org>

DOI: 10.1161/CIRCHEARTFAILURE.116.002973

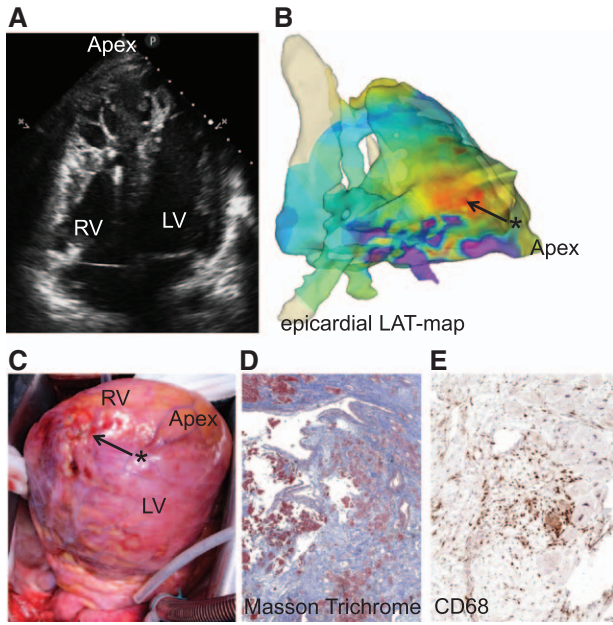


Figure. **A**, Echocardiographic 4-chamber view showing asymmetrical noncompaction cardiomyopathy. **B**, Epicardial electroanatomical map showing local activation time (LAT) during tachycardia. *Ablation site. **C**, Intraoperative situs. *Ablation site. **D** and **E**, Histology and immunohistology of excised myocardium show wide areas of fibrosis (Masson trichrome) and severe infiltration of CD68+ macrophages. LAT indicates local activation time; LV, left ventricle; and RV, right ventricle.

Successful Surgical Treatment of an Electrical Storm in a Patient With Atypical Noncompaction Cardiomyopathy

David Schibilsky, Karin Klingel, Jürgen Schreieck, Meinrad Gawaz, Tobias Walker, Christian Schlensak and Peter Seizer

Circ Heart Fail. 2016;9:

doi: 10.1161/CIRCHEARTFAILURE.116.002973

Circulation: Heart Failure is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231

Copyright © 2016 American Heart Association, Inc. All rights reserved.

Print ISSN: 1941-3289. Online ISSN: 1941-3297

The online version of this article, along with updated information and services, is located on the
World Wide Web at:

<http://circheartfailure.ahajournals.org/content/9/4/e002973>

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in *Circulation: Heart Failure* can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the [Permissions and Rights Question and Answer](#) document.

Reprints: Information about reprints can be found online at:
<http://www.lww.com/reprints>

Subscriptions: Information about subscribing to *Circulation: Heart Failure* is online at:
<http://circheartfailure.ahajournals.org/subscriptions/>