ACC22
APRIL 2 - 4 | WASHINGTON, DC

2514
JACC March 8, 2022
Volume 79, Issue 9, suppl A

Complex Clinical Cases

MANAGEMENT OF REFRACTORY SYMPTOMATIC PREMATURE VENTRICULAR CONTRACTIONS IN A PATIENT WITH LOUIS-DIETZ SYNDROME TYPE 3, EHLERS-DANLOS SYNDROME, AND SYSTEMIC LUPUS ERYTHEMATOSUS

Poster Contributions
For exact presentation time, refer to the online ACC.22 Program Planner at https://www.abstractsonline.com/pp8/#!/10461

Session Title: Complex Clinical Cases: FIT Flatboard Poster Selections -- Electrophysiology
Abstract Category: FIT: Electrophysiology

Authors: *Nolberto Jaramillo, Denis Malkov, Bernadette Riley, Todd J. Cohen, Seth I. Keller, New York Institute of Technology College of Osteopathic Medicine, Old Westbury, NY, USA, Mount Sinai Morningside Hospital, New York, NY, USA*

Background: Louis-Dietz syndrome (LDS) Type 3 is a rare disorder caused by an autosomal-dominant mutation in SMAD-3, altering the TGF-β pathway. LDS Type 3 typically manifests as aortic aneurysms and early-onset osteoarthritis, however other dermatologic, cardiovascular, and skeletal abnormalities have been reported.

Case: A 51-year-old woman was referred to the cardiology clinic for episodes of palpitations, syncope, chest pain, and shortness of breath during the COVID-19 pandemic. She had a history of congestive heart failure, cardiomyopathy, patent foramen ovale, atrial septal aneurysm, pre-COVID myocarditis, mitral valve prolapse, mitral regurgitation, and pericarditis. She also has a pertinent medical history of hypermobile Ehlers-Danlos syndrome (hEDS) and systemic lupus erythematosus (SLE). Her family and social history were remarkable for a daughter with SLE. Cardiopulmonary and general physical exams were remarkable for hypermobility. Evaluation with an ECG and Holter monitor showed normal sinus rhythm with unifocal premature ventricular contractions (PVCs) that correlated with her symptoms.

Decision-making: The patient was initially managed un-successfully with beta and calcium channel blockers. Cardiac ablation was subsequently performed on a left ventricular septal focus with remote magnetic navigation using the Niobe system from Stereotaxis inc. (due to its low risk for cardiac perforation). At 6 months follow up, the patient exhibited an increase in left ventricular ejection fraction from 40-50% to 55-60%, fewer symptoms, and fewer PVCs. She was later diagnosed with a right internal carotid artery aneurysm that prompted genetic testing that was positive for LDS Type 3.

Conclusion: This patient's unique combination of illnesses required a multidisciplinary team for management. The Stereotaxis robotic system safely and successfully treated the patient's PVCs and resulted in improvement of left ventricular function. Due to previous reports of arrhythmias associated with these connective tissue disorders, additional studies are necessary to understand the role of the SMAD-3 mutation, EDS, and SLE in contributing to arrhythmogenicity.