Mechanical and Electrophysiological Substrate for Recurrent Atrial Flutter Detected by Right Atrial Speckle Tracking Echocardiography and Electroanatomic Mapping in Myotonic Dystrophy Type 1

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A 48-year-old patient with myotonic dystrophy (MD) type 1, recurrent typical atrial flutter (AF), and otherwise unremarkable history was hospitalized for an electrophysiological study. The diagnosis of MD type 1 had been made 25 years earlier and was based on typical clinical features and confirmation by genetic analysis. The ECG pattern of AF was characterized by negative waves in the inferior leads and positive waves in V1, a cycle length of 288 milliseconds, and 2:1 atrioventricular conduction. At the time of the study, the patient was asymptomatic and showed normal findings at the cardiac physical examination, ECG, and standard echocardiography. Preablation ECG showed sinus rhythm at 60 bpm with a normal PR interval (180 milliseconds), regular QRS duration and morphology, and normal ventricular repolarization. Average septal-lateral mitral annulus velocities (s’, 9.5 cm/s; e’, 10.3 cm/s; a’, 7.4 cm/s), the E/e’ ratio (7.3), and left ventricular global longitudinal and circumferential strain (−21.5% and −23.2%, respectively) were all normal, as well as right ventricular systolic function (tricuspid annulus systolic excursion and peak systolic velocity, 25 mm and 18.5 cm/s, respectively). Analysis of longitudinal right atrial deformation by speckle tracking showed impaired strain mechanics in the inferior segment of the atrial septum (Figure 1, top, and Movie I in the online-only Data Supplement). Segmental atrial strain curves confirmed an abnormal deformation pattern at this level (Figure 1, bottom), resulting in reduced peak atrial longitudinal strain (average, 21.2%; normal value, >29.0%). Electroanatomic mapping of the right atrium in sinus rhythm (EnSite NavX System, Endocardial Solutions, St. Jude Medical Inc, St. Paul, MN) consistently showed a low-voltage area in the inferior portion of the atrial septum, suggesting regional scarring (Figure 2). Coronary sinus stimulation induced counterclockwise typical AF with the same ECG pattern of the clinical arrhythmia. Radiofrequency ablation of the cavotricuspid isthmus was performed, resulting in sinus rhythm restoration. Postprocedural activation mapping confirmed isthmus bidirectional block and success of the procedure (Figure 3). Postablation ECG was unchanged.

MD is an inherited multisystemic disease representing the most common autosomal-dominant muscular dystrophy worldwide. In its most severe form, called MD type 1 or Steinert disease, an abnormal expansion of a CTG-trinucleotide repeat sequence exists in the gene encoding for myotonic dystrophy protein kinase, a protein that plays a key role in intracellular signal modulation within smooth, cardiac, and skeletal myocytes. This mutation leads to progressive involvement of musculoskeletal apparatus, heart, brain, eye, endocrine, respiratory, and gastrointestinal systems. The typical neuromuscular pattern is characterized by progressive distal muscular atrophy and weakness, grip and percussion myotonia, ptosis, hatchet face, slurred speech, and rhinolalia, and it is often associated with multisystem manifestations such as cataracts, insulin resistance, dysphagia, oligospermia, and neurobehavioral disorders. Cardiac abnormalities are also commonly found in patients with MD type 1, including atrioventricular or intraventricular conduction defects, arrhythmias, left ventricular hypertrophy and dysfunction, heart failure, and sudden death. Magnetic resonance imaging studies and endomyocardial biopsies suggested that focal areas of myocardial fibrosis and fatty infiltration may be responsible for these disorders. Analysis of 2-dimensional strain by speckle tracking was recently proposed as a technique to discriminate between normal myocardium and scar tissue in other populations. However, its application in the study of atrial dynamics has not been tested in patients with MD type 1, and the existence of a relation between the segmental pattern of myocardial deformation and that of electrophysiological abnormalities in these patients was not assessed.

These images for the first time provide evidence of concordance between right atrial mechanical and electrophysiological abnormalities in a subject with MD type 1. Using an invasive electroanatomic mapping system, we found a segmental area of low voltages in the inferior portion of the atrial septum, suggesting regional scarring. High agreement between the segmental pattern of low-voltage myocardium and that of myocardial scarring on magnetic resonance imaging was previously established.
and recent data showed that electroanatomic mapping is even more sensitive than gadolinium-enhanced magnetic resonance imaging in detecting myocardial scars when tested against endomyocardial biopsy. Interestingly, the abnormal region identified by voltage mapping was concordant with the segmental impairment in 2-dimensional strain pattern detected by speckle tracking. Although a certain causal association between these abnormalities and the occurrence of relapsing AF cannot be established, these findings suggest that interesting future applications of speckle tracking—-to be tested in populations including patients with various types of AF—-might include noninvasive evaluation of potential arrhythmic substrate in the atrial myocardium; preprocedural assessment of segmental abnormalities in atrial mechanics, which may provide additional information about the potential electrophysiological mechanism underlying AF and the probable target site of ablation; and risk stratification for postablation arrhythmia recurrence.

**Figure 1.** Speckle tracking. **Top,** Color-coded visualization of right atrial (RA) 2-dimensional strain at ventricular end systole showing abnormal deformation in the inferior portion of the atrial septum (white arrow). Blue and red denote positive and negative deformation, respectively. **Bottom,** Strain curves confirming abnormal deformation pattern (white arrow) in the inferior segment of the atrial septum (red curve), with normal curves in the remaining segments (blue, superior atrial septum; pink, medial roof; green, lateral roof; turquoise, superior lateral wall; yellow, inferior lateral wall). AVC indicates aortic valve closure; LA, left atrium; LV, left ventricle; and RV, right ventricle.

**Disclosures**

None.

**References**

Figure 2. Electroanatomic mapping. Right ventricular voltage map obtained from the right posterior oblique (A) and left posterior oblique (B) views showing a low-voltage area (red-yellow-green-blue, <1 mV) in the inferior segment of atrial septum (yellow arrow). Normal myocardium is shown in violet. CS indicates coronary sinus; IVC, inferior vena cava; STL, septal tricuspid leaflet; and SVC, superior vena cava.

Figure 3. Electroanatomic mapping after radiofrequency ablation. Right atrial activation map obtained by coronary sinus stimulation from the right lateral (A) and right anterior oblique (B) views showing conduction block after successful cavotricuspid isthmus ablation. Normal activation is shown in red-yellow-green-blue; violet indicates late activation secondary to blocked conduction. IVC indicates inferior vena cava; STL, septal tricuspid leaflet; and SVC, superior vena cava.
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