Atrial Arrhythmias in Arrhythmogenic Cardiomyopathy: At the Beginning or at the End of the Disease Story? – Reply –

In a Letter to the Editor entitled “Atrial arrhythmias in arrhythmogenic cardiomyopathy – at the beginning or at the end of the disease story?” S. Peters describes two middle-aged female patients with morphological and electrocardiographic signs of arrhythmogenic right ventricular dysplasia (ARVD) who presented with atrial fibrillation (AF) as their first arrhythmic event. The author points to the observation that AF is very common in ARVD, with up to 40% of patients being affected, and may often precede ventricular tachyarrhythmias. On the other hand, Peters also indicates that in some patients AF may be a late consequence of progressed disease with severe right and left ventricular (LV) involvement, heart failure and atrial dilatation, which typically occur during the late phases of ARVD. Our study published in a previous issue of this Journal is in line with the letter by Peters showing that AF is common in our ARVD population (20%). As ARVD is a heterogeneous disease, the natural history may vary significantly among patients. Similar to the cases presented by Peters, AF occurred at a rather early stage of the disease in a minority of our study population. AF is common in patients with Brugada syndrome who do not have structural heart disease at the macroscopic level. As some forms of ARVD may overlap with the Brugada phenotype, and desmosomal mutations (plakophilin-2 and desmoglein-2) have been shown to impair cardiac sodium current (I Na), it is possible that associations (plakophilin-2 and desmoglein-2) have been shown to overlap with the Brugada phenotype, and desmosomal mutations within the desmosomal complex may affect ion channels, signal transduction and fibrofatty infiltration of the atria to a different extent than in the ventricles.

AF in ARVD is a very interesting topic, and future basic and clinical research is obviously necessary to better delineate and understand the various aspects of atrial arrhythmias in this challenging disease.

Disclosures
Sources of Funding: This work and the Zurich ARVC Program are supported by a grant from the Georg and Bertha Schwzyer-Winkler Foundation, Zurich, Switzerland.
Conflict of Interest: None declared.

References

Ardan M. Saguner, MD
Corinna Brunckhorst, MD
Department of Cardiology,
University Heart Center Zurich, Switzerland

Firat Duru, MD
Department of Cardiology,
University Heart Center Zurich;
Center for Integrative Human Physiology,
University Zurich, Switzerland

(Released online December 8, 2014)