APHRS–JHRS Joint Symposium	7月7日（土）／第3会場／9:00~11:00

Right Ventricular Cardiomyopathy in Asian Countries

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APJ-1

Genetic Background for Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia（ARVC/D）in Japan

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Arrhythmogenic right ventricular cardiomyopathy/dysplasia（ARVC/D）is an inherited heart muscle disease characterized by the fibrofatty replacement of right ventricle（RV）and is predominantly caused by mutations in genes encoding desmosomal proteins. ARVC/D often presents ventricular tachycardia（VT）of RV origin, heart failure and sudden death. We studied the genetic/clinical characteristics of Japanese ARVC/D patients.

Methods：The study included consecutive 35 Japanese probands（23 males）：29 with definite, 4 borderline and 2 possible diagnosis according to 2010 ARVC/D criteria. Four desmosomal protein genes were examined by direct sequencing methods：plakophilin-2（PKP2）, desmoplakin（DSP）, desmoglein-2（DSG2）, and desmocollin-2（DSC2）. **Results**：The mean age of first symptom in our patients was 38.6 ± 14.8 years, and mean age at diagnosis was 40.5 ± 17.7 years, while two were diagnosed without symptoms. We identified multiple desmosomal gene mutations in 19 probands：9 PKP2, 5 DSP, 2 DSG2, 1 DSC2, 1 PKP2 + DSP, and 1 DSP + DSG. As the first symptom, the cardiac arrest was significantly more frequent in young（＞40 years）patients（n = 5）than elder（＞40 years）group（n = 1）. PKP2 mutations were most frequently identified（n = 10, 28.6 %）. It should be noted that PKP2 mutations in younger probands were all premature stop codon, whereas only two probands carried stop codon mutations in elder probands（$p < 0.05$）.

Conclusions：We identified various mutations of desmosomal protein genes（19 of 35 patients（54 %）. The prevalence of the mutation carriers in the four genes was similar to that reported previously in Western countries. PKP2 mutations identified in young probands cause a premature stop codon, and their initial clinical manifestation was very severe. Therefore, the identification of genetically-affected family members, even though asymptomatic, would offer a strong clinical modality to prevent the cardiac sudden death.

APJ-2

Clinical Manifestations of Arrhythmogenic Right Ventricular Cardiomyopathy in Korean Patients

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Arrhythmogenic right ventricular cardiomyopathy（ARVC）is an important cause of syncope, ventricular arrhythmias, electrocardiogram（ECG）abnormalities and/or non-ischemic wall motion abnormalities.

Our previous postmortem study showed that ARVC is an important cause of sudden cardiac death in young（¡Â35 years old）Koreans. However, most of sudden cardiac deaths due to ARVC were not related to vigorous or competitive physical activity.

We also analyzed the clinical characteristics of living patients（44¡¾15 years old, 21 males）with ARVC. The common presenting symptoms were palpitations（32 %）, syncope/presyncope（24 %）and no symptoms（32%）. Four patients had a family history of premature sudden death or ARVC. Most patients with no symptoms were evaluated due to ECG abnormalities or asymptomatic ventricular arrhythmias. Ventricular tachycardia, ventricular fibrillation and frequent premature ventricular contractions only were observed in 41 %, 3 % and 22 %, respectively. Wall motion abnormalities of the right and left ventricles were detected in 92 % and 32 %, respectively. Fatty or fibrofatty infiltration was observed in most（≈80%）patients who underwent an endomyocardial biopsy. Two patients had signs of heart failure. Two patients with syncope/presyncope were diagnosed with vasovagal syncope and another was due to side effects from a medication. Most of the patients with ventricular arrhythmias were treated with β-blockers and/or amiodarone. Implantable cardioverter-defibrillator（ICD）was implanted in 3 patients. During a mean follow-up of 4 years no syncope or sudden death developed except for in one patient with an ICD who suffered from recurrent shocks due to ventricular fibrillation.

ARVC may be an important cause of syncope, ventricular arrhythmias, and ECG and wall motion abnormalities of the ventricles in Koreans. Some patients, however, do not have a typical clinical presentation. Thus, a high clinical suspicion and extensive studies may be needed to establish the diagnosis of ARVC.

APJ-3

Radiofrequency Catheter Ablation of Ventricular Tachycardias in Arrhythmogenic Right Ventricular Cardiomyopathy（ARVC）

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Arrhythmogenic right ventricular cardiomyopathy（ARVC）is a major cause of sudden cardiac death and ventricular tachyarrhythmias（VT）in relatively young, healthy individuals.
Patients at higher risk should undergo ICD placement for primary and secondary prevention. However, the results from long-term follow up confirmed the necessity of ICD therapy and also the progressivly high failure rate of the devices.
Although there is demand for catheter ablation-especially when cultural and/or economic reasons are considered, the concerns of the progressive nature of ARVC substrate, the limited number of most studies and the poorly defined mapping and ablation strategies make such procedure challenging.
During the last decade, with the guidance of 3D electroanatomic mapping techniques, various tools have been applied for targeting VT foci：substrate mapping（combined with entrainment if possible）with linear ablation block the potential conduction channel, ablation of delayed isolated potential, substrate and pace mapping for fast or unstable VTs and noncontact mapping with regional ablation. These studies have reported varied outcomes with long-term succes rate ranged from 15%～82%.
More recently, the epicardial approach was applied after the failure in endocardial, the results seemed satisfyingand consistent. However, we have archived a relatively high success rate in a large cohort of ARVC patients（more than 80 pts. 40% with history of syncope）with endocardial approach only. We own this to the unipolar noncontact array mapping, extensive stimulation protocol of VT induction and regional ablation strategy.
Although the recurrence rates remain considerable even if the endo-and epi-cardial approaches were both applied, the results of catheter ablation of ARVC VTs have been improving significantly and it seems more and more expecting.

APJ-4

Predictors of Cardiac Events in Right Ventricular Cardiomyopathy：a Report from a Long-Term Observational Study of 74 Asian Cases

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Background：Arrhythmogenic right ventricular（RV）cardiomyopathy（ARVC）is known as a cardiomyopathy characterized by life-threatening ventricular arrhythmias and congestive heart failure because of ventricular dysfunction of the RV and/or left ventricle（LV）involvement. The aims of this study were to examine the clinical presentation and to identify long-term prognostic predictors in a large cohort of patients with ARVC.
Methods：74 patients with ARVC over 1 year follow-up（Male 61, Aged 46±14 yrs）were enrolled. Primary study endpoints were death and hospitalization for heart failure.
Results：During mean follow-up period of 10±7 years, 70 patients had a history of ventricular tachycardia, and ICD was implanted in 20 patients. Hospitalization for heart failure（HF）was documented in 15 patients（20%）, 6 patients were died from sudden cardiac death and 3 from HF. In 18 patients（24%）, Atrial tachyarrhythias were developed（atrial tachycardia：6, atrial flutter：5, atrial fibrillation：10）. Kaplan-Meier's curve showed that ARVC patients with ATs had the greater risk of HF hospitalization（p=0.0036）and death from HF（p=0.031）. Multivariate analysis showed that ATs was independent predictor of HF in ARVC.（HR 10.0, 95% CI 0.46-1.92, p=0.0017）. Three autopsy cases died from heart failure revealed that fibro-fatty replacement was also identified in right atrium, as well as right ventricle.
Conclusions：Atrial arrhythmias were developed in the patients with ARVC during long-term follow-up and they can be aggravating factor of heart failure.