Introduction: Early repolarization syndrome (ERS) is generally diagnosed in patients who display ER pattern in the inferior and/or lateral leads presenting with aborted cardiac arrest, documented VF, or polymorphic VT. In J-wave syndromes expert consensus conference report endorsed by the APHRS (J Arrhythm, 2016), Shanghai Score System was proposed for diagnosis of ERS. However, it remains unclear whether the scoring system is truly effective in predicting VF occurrence in subjects displaying ER pattern. In the present study, therefore, we retrospectively evaluated the validity of Shanghai Scoring System in our Japanese ERS patients.

Methods: Thirteen consecutive Japanese ERS patients (mean age 46 ± 17 years) with a history of spontaneous VF, who admitted to Oita University Hospital between 2005 and 2018, were retrospectively evaluated. All the patients were implanted with implantable cardioverter-defibrillators (ICDs). The patients were scored according to the Shanghai Score System.

Result: During the follow-up period of 50.0 ± 45.2 months (mean ± SD), three patients experienced appropriate shock delivery due to the recurrence of VF after ICD implantation (recurrent VF group). The VF recurrence was observed 11.1 ± 8.7 months after the ICD implantation. In the recurrent VF group, one patient showed augmented amplitude of J-waves with horizontal ST-segment while the other 2 patients showed dynamic changes in J-wave amplitude. In intracardiac electrograms of ICD recordings, short-coupled VPCs invariably preceded the development of VF in the recurrent VF groups. The total points of Shanghai Score System in the recurrent VF group were significantly higher than those in the non-recurrent VF group (6.7 ± 0.2 vs 4.4 ± 0.6 points, P<0.001).

Conclusion: Results of our Japanese small-scale study suggest that Shanghai Scoring System can effectively identify the patients at high risk for VF recurrence in ERS patients. Its usefulness in subjects with ER pattern who do not have a history of lethal ventricular tachyarrhythmia should be further evaluated.