## Cardiac function at diagnosis is important prognostic factor in patients with cardiac sarcoidosis -from Japanese nationwide questionnaire survey-

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**Background:** Sarcoidosis is a systemic non-caseating granulomatous disease of unknown etiology. Cardiac involvement (cardiac sarcoidosis, CS) has been reported to be an important prognostic factor in this disease because of heart failure and/or ventricular arrhythmia, and corticosteroid therapy is usually prescribed to prevent cardiac events. However, little is known about the relationship of cardiac function and concomitant corticosteroid therapy on later cardiac events in CS.

**Objective:** We evaluated the relationship between prognosis and left ventricular ejection fraction (LVEF) at the time of diagnosis in CS patients from the Japanese nationwide questionnaire survey.

**Methods:** Total of 757 Japanese patients from 57 hospitals who diagnosed CS were examined. Patients who unsatisfied the criteria of the Japanese new guidelines, or who underwent cardiac transplantations were excluded, and 420 patients (287 females, mean age 60±13 years old, median follow-up periods 1864 days [interquartile range: 845–3159 days]) were analyzed. The relationship of adverse events (all-cause death, cardiovascular death, and appropriate ICD [Implantable Cardioverter Defibrillator] discharge) and

LVEF (with corticosteroid 84%) (low LVEF: LVEF $\leq$ 35% n=98 [with corticosteroid in 78%], moderate LVEF: LVEF 35–50% n=104 [with corticosteroid in 93%], normal LVEF: 50 $\leq$ LVEF n=218 [with corticosteroid in 83%]) were evaluated respectively.

**Results:** 89 CS patients developed all-cause death (n=50), cardiovascular death (n=30) or appropriate ICD discharge (n=48). The frequency of corticosteroid therapy was not different in the each LVEF group, but Kaplan-Meier analysis revealed that all-cause death, cardiovascular death, and all cardiovascular adverse events were more observed in lower LVEF group (log-rank p<0.0001). Furthermore, multivariate Cox hazard analysis revealed that LVEF was a most important independent prognostic factor in CS.

**Conclusion:** This Japanese nationwide questionnaire survey data showed that initial LVEF was an independent and strong prognostic predictor in CS, therefore primary prevention would be needed even after starting corticosteroid in patients with decreased cardiac function.

Kaplan-Meier curves for all-cause death, cardiovascular events in cardiac sarcoidosis



All cause death(A), Cardiovascular death(B) and Cardiovascular death and ICD discharge(C) occurred in patients lower LVEF groups compared with normal LVEF groups (log-rank p<0.0001)