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Title: Incidence, clinical manifestations and prognosis of cardiac sarcoidosis in patients with pulmonary sarcoidosis

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Body: Background Early diagnosis of cardiac sarcoidosis (CS) is vitally important in the management of patients with pulmonary sarcoidosis (PS), because of the risk of sudden cardiac death. Some patients with PS develop CS during follow-up, but few studies have addressed its incidence and clinical course. Aim To investigate the incidence, initial manifestations, and prognosis of CS in patients with PS. Methods Participants comprised 215 subjects who had been pathologically diagnosed with PS since January 2002. The incidence of CS was evaluated using the diagnostic guidelines for CS of the Japan Society of Sarcoidosis. 18F-fluoro-2-deoxyglucose positron emission tomography and cardiac magnetic resonance imaging were included in the diagnostic work-up unless contraindicated. Initial manifestations and prognosis of CS were also reviewed. Results Twenty (9.5%) of the 215 PS patients developed CS during follow-up (median, 2.5 years; range, 0.8-4.5 years). Annual incidence rate of CS was 3.8%. Initial manifestations of CS were abnormal findings on screening electrocardiogram (n=12), chest symptoms such as palpitation (n=6), and others (n=2). During the follow-up of CS (median, 2.6 years; range, 0.4-4.6 years), three died (lung cancer, n=2; sudden cardiac death, n=1) and four were implanted with a permanent pacemaker or implantable cardioverter-defibrillator. Conclusions CS developed in PS patients at an annual incidence rate of 3.8%, with electrocardiogram changes and chest symptoms being the most prevalent initial manifestations. Of the 20 CS patients, 25% encountered CS-related death or were forced to be treated by device implantation during the follow-up.