

## **Abstract**

**Background:** Amyloidosis is a rare disease caused by tissue deposition of protein fibrils. In case of cardiac involvement, it is called cardiac amyloidosis (CA), which has a poor prognosis and causes sudden cardiac death (SCD). An implantable cardioverter defibrillator (ICD) prevents SCD due to ventricular tachyarrhythmia; however, ICD implantation in patients with CA is controversial because of the poor prognosis of CA and the possibility of pulseless electrical activity. This study is a systematic review aimed to evaluate the efficacy of ICD in patients with CA.

**Methods:** A systematic literature review was conducted in compliance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines (PRISMA). On June 30, 2019, a comprehensive literature search of Medline, PubMed, Embase, CINAHL and the Cochrane Library was performed. Longitudinal observational studies of patients with CA with implanted ICD were included. Primary outcomes were appropriate ICD therapy, all-cause mortality and one-year survival.

**Results:** Nine studies with a total of 704 CA patients with implanted ICD were included. The median incidence rate of appropriate ICD therapy was 26.0% (range 6.5-28.3%) of the patients during 21.5 (0-175) months of follow-up period. All-cause mortality was observed in 96 (41.3%) among 232 patients and median one-year survival was 79% (range 39-92%).

**Conclusions:** Patients with CA benefit from ICD implantation considering the incidence rate of appropriate ICD therapy and one-year survival. There is no evidence that ICD implantation can

improve patients' prognosis. Larger studies are needed to elucidate the impact of ICD on patients with CA and to develop selection criteria for implantation.

## **1. Introduction**

Amyloidosis is a rare systemic disease caused by tissue deposition of protein fibrils, and it reduces the function of various organs.<sup>1</sup> Amyloidosis is classified by the protein component into immunoglobulin light chain (AL) amyloidosis, transthyretin related (ATTR) amyloidosis, serum amyloid A (AA) amyloidosis, and isolated amyloidosis.<sup>1,2</sup> The incidence rate per million person-years of AL amyloidosis, which is the most common type of amyloidosis, was reported to be 10.5.<sup>2</sup> In case of cardiac involvement, it is called cardiac amyloidosis (CA), which is related to a poor prognosis. The life expectancy of patients with AL amyloidosis and heart failure is reported to be less than one year,<sup>3</sup> although the prognosis of CA depends on the type of amyloidosis. The majority of cases of CA is caused by AL amyloidosis and ATTR amyloidosis among the various types of amyloidosis.<sup>4</sup>

It is reported that CA is related to various types of arrhythmia including ventricular tachyarrhythmia, atrial tachyarrhythmia and bradyarrhythmia,<sup>5</sup> and two thirds of patients with CA die of cardiac cause and half of them experience sudden cardiac death (SCD).<sup>6,7</sup> Implantable cardioverter defibrillator (ICD) is useful to prevent SCD owing to ventricular tachycardia (VT) or ventricular defibrillation (VF). There are some reports of successful defibrillation by ICD in patients with CA.<sup>8</sup>