Cardiac Involvement in Emery-Dreifuss Muscular Dystrophy and Related Management Strategies.

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Abstract
Emery-Dreifuss muscular dystrophy (EDMD) is a group of hereditary muscular dystrophy syndrome caused by deficiency of genes encoding nuclear envelope proteins. Patients having EDMD show the triad of muscle dystrophy, joint contracture, and cardiac disease. In almost all patients, cardiac involvement is prevalent and is the most severe aspect of EDMD. Cardiac disease is predominantly shown by conduction defects, atrial fibrillation/flutter, and atrial standstill. Sudden death and heart failure because of left ventricular dysfunction are important causes of mortality, particularly in those patients that have the LMNA mutation. Medical treatment of EDMD is limited to addressing symptoms and ambulation support; moreover, pacemaker implantation is necessary when there are severe conduction defects and bradycardia occurs. Note that automated defibrillation devices may be considered for those patients who have a high risk of sudden death, rate, or rhythm control. Also, anticoagulation should be initiated in those patients who have atrial fibrillation/flutter. Thus, for optimal management, a multidisciplinary approach is required.

KEYWORDS: Arrhythmia; Skeletal muscle disease

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