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Cardiac complications of Beta-thallesemia

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Beta-thalassemias are a group of inherited, autosomal recessive diseases, characterized by reduced or absent synthesis of betaglobin chains of the hemoglobin tetramer, resulting in variable phenotypes, ranging from clinically asymptomatic individuals to severe anemia. Cardiac complications represent a leading cause of mortality in β-TM patients, although an important and progressive increase of life expectancy has been demonstrated after the introduction of chelating therapies. Iron overload is the primary factor of cardiac damage resulting in thalassemic cardiomyopathy, in which diastolic dysfunction usually happens before systolic impairment and overt heart failure (HF). Although iron-induced cardiomyopathy is slowly progressive and it usually takes several decades for clinical and laboratory features of cardiac dysfunction to manifest,arrhythmias or sudden death may be present without signs of cardiac disease and only if myocardial siderosis is present. Careful analysis of electrocardiograms and other diagnostic tools may help in early identification of high-risk β-TM patients for arrhythmias and sudden cardiac death.

Despite the advances in the management of β-TM, heart disease remains the leading cause of mortality in these patients. Cardiac arrhythmias are frequent in β-TM patients, particularly in the advanced stage of the disease, when a significant cardiac iron loading is present.)e cardiovascular evaluation of β-TM patients should be performed by cardiologists, with experience in clinical arrhythmology and echocardiography, who have knowledge of thalassemia and

iron-related cardiotoxicity.)e ECG analysis should include the measuremtent of P wave and QT interval dispersion; the echocardiogram should include the evaluation of the atrial electromechanical delay or left atrial function analysis.

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