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Electrocardiogram's Particularities in Duchenne Muscular Dystrophy

Abdallah Fayssoil

CHU Raymond Poincare, APHP, Garches, France

Abstract

Duchenne muscular dystrophy is a genetic neuromuscular disease that affects young patients. Cardiac involvement is classical in this disease. Electrocardiogram discloses pattern particularities that are described in this paper.

Keywords: Duchenne muscular dystrophy; Dystrophin; Heart; ECG

Duchenne muscular dystrophy (DMD) is an X-linked recessive disorder, caused by the absence of dystrophin, which affects 1 of 3.500 male births [1]. Dystrophin is a protein located on the inner side of skeletal and cardiac muscle cells. This protein links cytoskeleton to the extra cellular matrix. The lack of dystrophin leads to progressive fiber degeneration. The consequence is a progressive muscle wasting and weakness of variable distribution. Clinical manifestations of DMD are visible before the age of 6 years and are characterized by progressive loss of strength. By the age of 12, most patients are confined to wheelchair. Cardiac involvement is present in about 90% of the patients and 20% of patients die from cardiac complications [1]. Heart involvement is due to progressive replacement of cardiomyocytes and Purkinje system by connective tissue or fat [1]. Electrocardiogram (ECG) abnormalities include tall R waves in leads V1 V2 (R/S ratio>1), abnormal Q-waves in lateral leads. Sinus tachycardia, shortened PQ interval, prolonged QT interval, atrial premature, supra-ventricular arrhythmias, ventricular premature beats and conduction abnormalities are also reported [2]. Sinus tachycardia is the most frequent cardiac abnormality. It has been reported at least 2 asymptomatic premature ventricular beats in 33% of young DMD patients (age from 12 to 24 years) [3]. Tachycardia may be attributed to cardiac autonomic nervous impairment [4]. Arrhythmias are attributed to progressive fibrosis of the cardiac conduction system and impairment in the cardiac autonomous nervous system [1]. Atrophy and scaring of the postero-basal region and the adjacent lateral wall of the left ventricle are reported and may explain the ECG pattern with abnormal Q-waves. In DMD, there is paucity of symptoms and abnormal physical findings of cardiac disease despite evidence in some of significant cardiac involvement. Echocardiography is essential for evaluation of cardiac dysfunction.

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***Corresponding authors:** Abdallah Fayssoil, CHU Raymond Poincare, APHP, Garches, France, E-mail: fayssoil2000@yahoo.fr

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