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X Marks the Spot: Duchenne's Cardiomyopathy

To the Editor:

Dilated cardiomyopathy is a common finding in individuals with Duchenne's muscular dystrophy, being present in virtually all patients over 18 years of age.[1] We describe a case of Duchenne's cardiomyopathy with classic electrocardiogram findings.

An 18-year-old male patient with a history of Duchenne's muscular dystrophy, secondary restrictive lung disease, and cardiomyopathy presented with pneumonia. An electrocardiogram (ECG) (**Figure**) on admission demonstrated sinus tachycardia with a ventricular rate of 108 beats per minute, right axis deviation, tall R wave in V1, delayed R wave progression in precordial leads, and Q waves in II and V4-V6. These ECG findings were consistent with Duchenne's cardiomyopathy. A transthoracic echocardiogram showed a mildly dilated left ventricle, severe diffuse hypokinesis, and a left ventricular ejection fraction of 15%-20%, decreased from his baseline of 35%.

Common ECG changes in Duchenne's include short PR interval, tall R wave with an abnormal R/S ratio in V1, and deep Q waves, most frequently in V5 and V6.[2] Other findings include Q waves in inferior leads, right ventricular hypertrophy, and T-wave inversions and ST-segment depression in inferolateral leads. Perloff et al[3] in 1966 proposed that preferential myofilament loss in the posterobasal segment leads to the decrease in posteriorly directed forces and an increase in anterior forces, resulting in a tall R wave and abnormal R/S ratio in V1. More recent cardiac magnetic resonance imaging studies have shown frequent involvement of the lateral segment as well.[4]

There is debate about the significance of these ECG findings. Most studies suggest that presence of ECG

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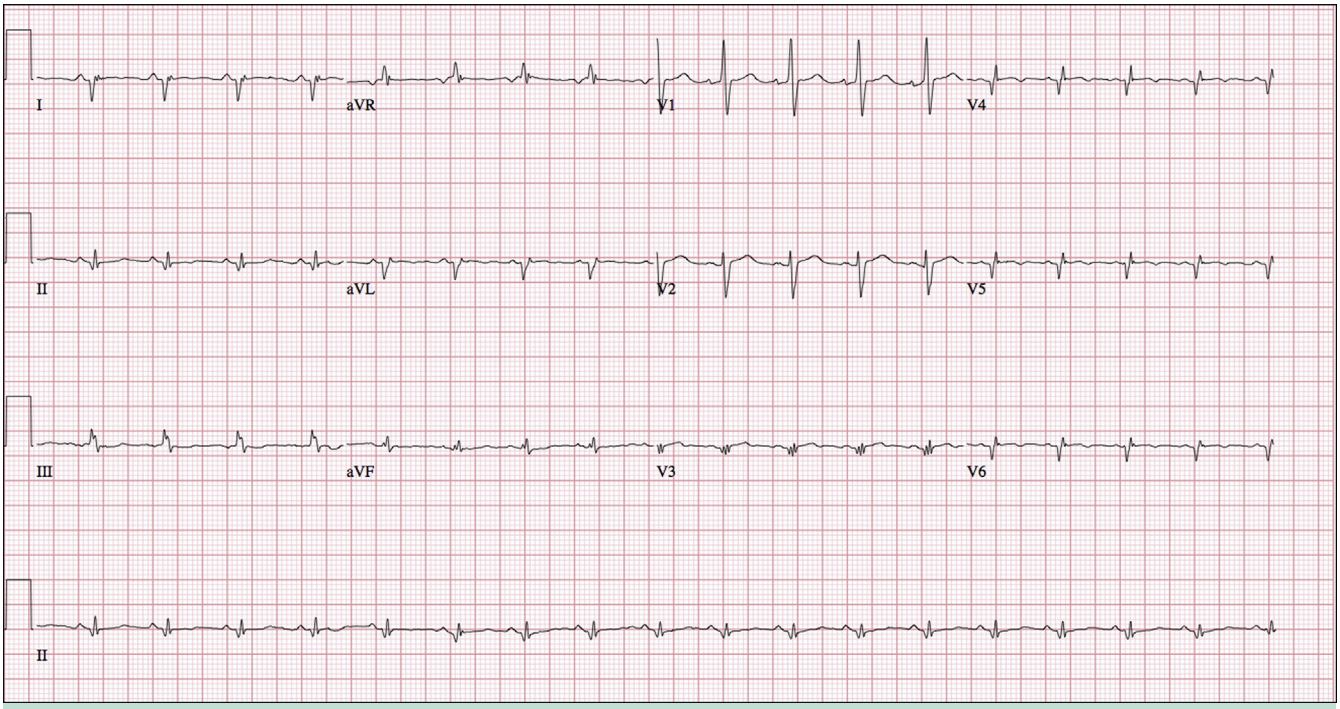

Figure Electrocardiogram. Sinus tachycardia, right axis deviation, tall R wave in V1, delayed R wave progression in precordial leads, and Q waves in II and V4-V6.

changes is unrelated to age, disease severity, or cardiac function, while others found sequential ECG changes correlating with disease progression.[5] Thrush et al[6] found that there was no significant difference in the prevalence of ECG findings between Duchenne's patients with and without cardiomyopathy, so it is possible that ECG findings reflect cardiac changes related to dystrophin deficiency rather than clinically apparent cardiomyopathy. Furthermore, there is no correlation between types of mutations and ECG abnormalities, again supporting the idea that ECG findings reflect general deficits in dystrophin.[7]

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