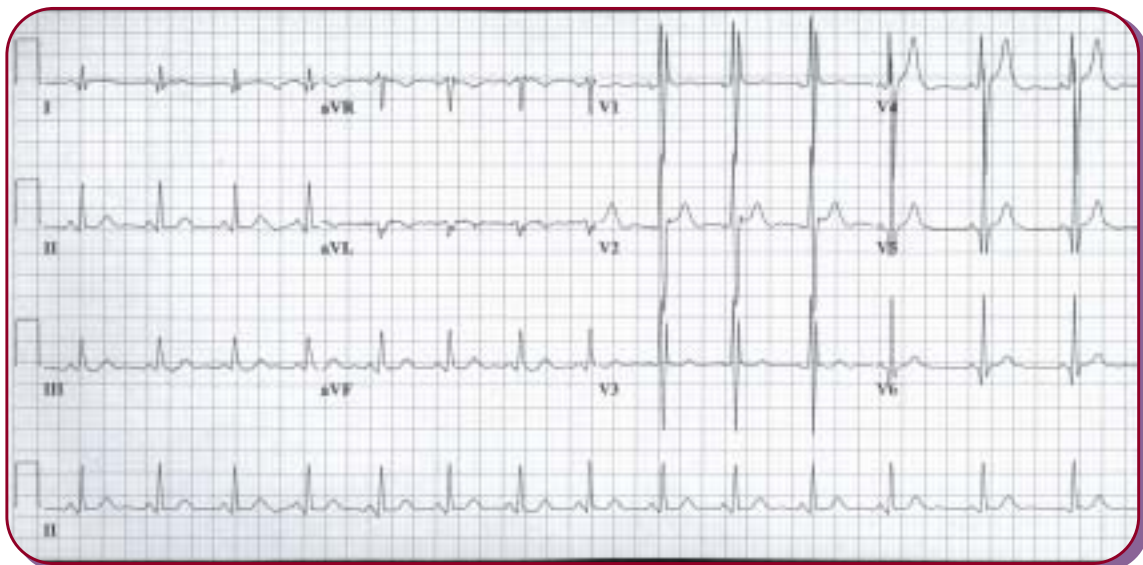


An Unusual Adolescent ECG

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This echocardiogram was recorded in an 18-year-old male who presented to the emergency department complaining of atypical chest pain. His total creatine kinase (CK) and CK-MB iso-enzyme levels were moderately elevated.

1. What ECG abnormalities are present?
2. What is the most likely diagnosis?



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This Month's ECG Diagnosis

1. The rhythm is sinus with a relatively short PR interval of 100 ms. The heart rate slows in the latter portion of the recording, perhaps due to sinus arrhythmia. The most unusual features are the large R waves in leads V₁ and V₂ and the presence of relatively deep, but narrow, Q waves in leads V₅ and V₆. Q waves are also present in leads I and aVL and there is an incomplete right bundle branch block (RBBB) pattern consistent with a right ventricular conduction delay.

2. The ECG is typical of Duchenne muscular dystrophy. This X chromosome-linked, recessive condition is associated with a unique form of myocardial dystrophy confined to the posterobasal portion of the free wall of the left ventricle. Myocyte degeneration and replacement by fibrous tissue lead to loss of normal electrical forces and ECG changes, which might be misinterpreted as being due to a posterior wall myocardial infarction (MI).

Abnormalities of cardiac rhythm and/or conduction are well described in Duchenne muscular dystrophy and include:

- short PR intervals due to accelerated atrioventricular node conduction,
- intraventricular conduction disturbances (usually right ventricular conduction delays manifesting as incomplete RBBB), and
- atrial and ventricular arrhythmias.

Marked heart rate variability and exaggerated sinus arrhythmia are unusual; whether this is due to autonomic dysfunction is unclear.

Were it not for the young age and clinically apparent diagnosis in this patient, the abnormal ECG and elevated CK levels might have raised concern about the possibility of recent MI. However, elevations of circulating CK are invariable in Duchenne patients and, due to abnormal expression of this isoenzyme from dystrophic skeletal muscle, elevated CK-MB levels are also commonly seen. 