# Is this familial hypertrophic cardiomyopathy?

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*ECG Education articles are inspired by, but not based on, real cases to illustrate the importance of knowledge about ECGs in relation to clinical situations in general practice. Management is not discussed.*

**Jamie is 19 years old. His older sister has recently been diagnosed with apical variant hypertrophic cardiomyopathy. Jamie is very fit and plays competition rugby every weekend. He and his mother present today because Jamie needs a referral to a cardiologist for further investigation as he has been told this condition is hereditary. His mother has already been tested and her ECG is normal. Jamie is taking no medication and has had no medical problems in the past. You arrange an ECG for Jamie before he sees the cardiologist.**

### Q1. What does the patient's ECG show (see below)?

The ECG shown below shows precordial and lateral T-wave inversions. Note also that the P waves are large and bifid, suggesting left atrial enlargement due to raised left ventricular filling pressures.

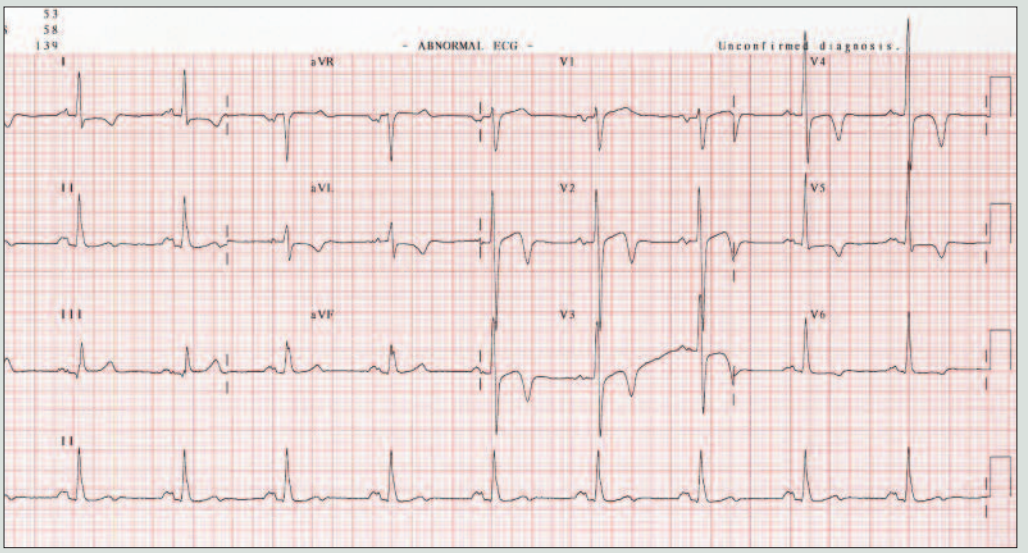
### Q2. What does the T wave represent?

On an ECG, the T wave represents repolarisation of the ventricles. The T wave is normally upright in leads I, II and V3 to V6 and normally inverted in aVR and sometimes V1. T waves are variable but also usually upright in leads III, aVL, aVF and V2. They are usually 1 to 5 mm in amplitude in leads I, II and III and 2 to 10 mm in leads V1 to V6. Their duration is not normally measured. The duration of the T wave contributes substantially to the QT interval (which also includes the QRS duration and ST segment).

### Q3. What is T-wave inversion associated with?

T-wave inversion is associated with the following.

- Coronary ischaemia (acute or recent).
- Subendocardial myocardial infarction – the infarction is not full thickness and there is T-wave inversion but no Q waves.
- Critical proximal left anterior descending artery stenosis in association with unstable angina (Wellens' sign) – deep T-wave inversion (sometimes biphasic) over the precordial leads V2 to V3.
- Left ventricular hypertrophy – there may be T-wave inversion in V5, V6, II and aVL.
- Right ventricular hypertrophy – there may be T-wave inversion in V1, V2 and V3.
- Use of digoxin is associated with sloping depression of the ST segment; therefore, an ECG should be carried out as a baseline before prescribing this medication.
- Aberrant ventricular conduction, such as bundle branch blocks, pre-excitation (through accessory pathways, as seen in Wolff-Parkinson-White syndrome), ventricular ectopics and electronic ventricular pacing.
- T-wave memory – transient T-wave inversion (lasting hours to weeks) after resolution or removal of aberrant ventricular conduction, such as bundle branch block, pre-excitation, pacing or frequent ventricular ectopics.
- Pulmonary embolism – typically shallow inferior lead inversion, but if associated with right ventricular strain, deeper T-wave

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inversion is seen in the right to mid precordial leads.
- Persistent juvenile T-wave pattern – T-wave inversion is normal in leads V1 to V3 in children.
- Severe cerebral damage, subarachnoid haemorrhage and status epilepticus.

### Q4. Can T-wave inversion be normal?

T-wave inversion can be normal in the following cases.

- T-wave inversion in leads V1 to V3 is normal in children.
- T-wave inversion in lead III may be a normal variant. New T-wave changes, however, are generally not. Variations in ECG electrode placement can sometimes cause spurious new T-wave changes.
- Generally, T-wave inversion over 2 mm in two or more adjacent leads requires further investigation. Inferior or lateral lead T-wave inversion should also be followed up in case of apical variant hypertrophic cardiomyopathy.
- T-wave inversion with normal cardiac imaging and cardiac enzymes occurs more commonly in fit women and men of native African origin than in Caucasians. Even so, it is recommended that any individuals with T-wave inversion in leads other than aVR and V1 should have annual ECGs, cardiological review and/or echocardiography in case of an evolving cardiomyopathy.

### Q5. What other changes in the T wave can there be?

Other changes in the T wave include the following.

- Transmural myocardial infarction produces ST elevation.
- Flat T waves (under 1 mV in the limb leads and 2 mV in the precordial leads) are associated with hypokalaemia, ischaemia and low QRS voltage (which may be due to cardiomyopathy, previous infarction or obesity).
- Tall or peaked T waves with a narrow base are associated with hyperkalaemia.
- Tall T waves with a broad base may be due to very early ST elevation myocardial infarction and also Prinzmetal's angina.
- Biphasic T waves occur in myocardial ischaemia and hypokalaemia.
- T-wave alternans is a beat-to-beat variation in the amplitude or shape of the T wave. It is associated with lethal ventricular tachycardias.
- Double peaked ('camel hump') T waves occur when the U wave is incorporated into the T wave or when the P wave is hidden in the T wave (as occurs with supraventricular tachycardias and some AV blocks).

### Q6. What is apical variant hypertrophic cardiomyopathy?

This is a hereditary disease of the cardiac muscle that mainly affects the left ventricular apex. It is most common in Japanese people, often occurs in others from Asian countries and is relatively rare in Caucasians. It is thought to have a generally better prognosis than some other forms of hypertrophic cardiomyopathy, but is associated with sudden cardiac death, malignant arrhythmias, apical myocardial infarctions and aneurysms. Characteristically, the cardiac echocardiography shows a 'spade shape' of the left ventricular space at end diastole. Well-established cases show a typical giant T wave negativity, especially in the inferior and lateral leads.

### Outcome

*You call Jamie and his mother back for another appointment because the ECG is not normal. You discuss the results of the ECG with them and arrange an early appointment for Jamie to see the cardiologist (as opposed to the usual waiting time of two months plus). In the meantime, you advise Jamie not to play football until he has been reviewed by the cardiologist.*

*The cardiac echocardiography carried out by the specialist was inconclusive, and so a cardiac MRI was arranged. This showed localised hypertrophy of the apical region. Given the family history, this result suggests Jamie may have the same condition as his sister and will need specialist assessment in the future. In light of this, Jamie has decided not to pursue competition rugby as his future career. Jamie's father should also be tested for the condition.* **CT**

## Key points

- **The T wave is normally upright in leads I, II and V3 to V6 and normally inverted in aVR and sometimes V1.**
- **T waves are variable but usually upright in leads III, aVL, aVF and V2.**
- **T waves are usually 1 to 5 mm in amplitude in leads I, II, and III and 2 to 10 mm in leads V1 to V6.**
- **T-wave inversion in leads V1 to V3 is normal in children.**
- **Generally, T-wave inversion over 2 mm in two or more adjacent leads requires further investigation.**