Detection of apical hypertrophic cardiomyopathy by cardiovascular magnetic resonance in patients with non-diagnostic echocardiography

J C C Moon, N G Fisher, W J McKenna, D J Pennell

Objective: To investigate the role of cardiovascular magnetic resonance (CMR) in a series of patients with ECG repolarisation changes and normal echocardiography.

Patients and design: 10 patients with anterolateral T wave inversion for which there was no obvious pathological cause who had normal routine echocardiography without contrast for the exclusion of hypertrophic cardiomyopathy (HCM) also had CMR that was diagnostic of apical HCM.

Results: Apical HCM detected by CMR could be morphologically severe with wall thickness up to 28 mm, or mild. The extent of repolarisation abnormalities did not correlate to the morphological severity.

Conclusions: In patients with unexplained repolarisation abnormalities, a normal routine echocardiogram without contrast does not exclude apical HCM. Further imaging with CMR or contrast echocardiography may be required. The reliance on routine echocardiography to exclude apical HCM may have led to underreporting of this condition.

METHODS

Patients

The 10 patients (eight men, mean (SD) age 56 (5) years) all had repolarisation changes in their anterolateral leads in the form of T wave inversion, three with giant negative T waves. In all cases a pathological cause for the electrocardiographic changes had not been determined but HCM was suspected. Five patients had been found to have an abnormal ECG on routine medical examination. Four had unexplained symptoms, of whom one had a family history of an abnormal ECG and one was found because of a family history of coronary artery disease. None of the patients had a history of hypertension but one was an amateur long distance runner. No patient had a family history of HCM. Table 1 presents patient details and results.

Echocardiography

All patients had undergone an echocardiography investigation at either Royal Brompton Hospital or St George’s Hospital, London. In all cases the presence of apical HCM was not confirmed. The echocardiography machines used were the Hewlett-Packard Sonos 5500 system with S4 probe or GE system fVe. Both machines have harmonics available. Views were reported as poor for one patient and appeared poor on review for one other patient. No patient at this stage underwent contrast echocardiography.

CMR

CMR was performed without the use of an exogenous contrast agent on a 1.5 T Siemens Sonata scanner (Erlangen, Germany) with front and back surface coils and prospective electrocardiographic triggering. Imaging consisted of scouts and subsequent two and four chamber fast prospective ECG triggering.

Image parameters were a repetition time of 3.2 ms, echo time 1.6 ms, in-plane pixel size 2.3 x 1.4 mm, slice thickness 7 mm, and flip angle 60°, acquired in about 12 heart beats.

The diagnosis of apical HCM with spade like configuration was based on published criteria of both an apical wall thickness > 15 mm and a ratio ≥ 1.3 of maximum left ventricular short axis thickness at the apical level to the basal.
Table 1  Patient characteristics and results from the 10 study patients, all of whom had cavity obliteration on cardiovascular magnetic resonance (CMR)

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Reason for ECG</th>
<th>Depth of T wave inversion (mm)</th>
<th>Maximum apical thickness by CMR (mm)</th>
<th>Average apical basal ratio by CMR</th>
<th>Echocardiography report</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>48</td>
<td>M</td>
<td>Family history of IHD</td>
<td>11</td>
<td>27</td>
<td>1.8</td>
<td>Normal</td>
</tr>
<tr>
<td>2</td>
<td>51</td>
<td>M</td>
<td>Routine medical examination</td>
<td>12</td>
<td>16</td>
<td>1.4</td>
<td>Trabeculated apex</td>
</tr>
<tr>
<td>3</td>
<td>64</td>
<td>F</td>
<td>Palpitations, chest pain</td>
<td>17</td>
<td>28</td>
<td>3.2</td>
<td>Akinetic apex</td>
</tr>
<tr>
<td>4</td>
<td>56</td>
<td>M</td>
<td>Chest pain</td>
<td>6</td>
<td>15</td>
<td>1.4</td>
<td>Normal</td>
</tr>
<tr>
<td>5</td>
<td>57</td>
<td>M</td>
<td>Family history of abnormal ECG</td>
<td>4</td>
<td>16</td>
<td>2.0</td>
<td>Normal</td>
</tr>
<tr>
<td>6</td>
<td>56</td>
<td>M</td>
<td>Routine medical examination</td>
<td>11</td>
<td>20</td>
<td>2.5</td>
<td>Normal</td>
</tr>
<tr>
<td>7</td>
<td>59</td>
<td>M</td>
<td>Routine medical examination</td>
<td>10</td>
<td>16</td>
<td>2.5</td>
<td>Normal</td>
</tr>
<tr>
<td>8</td>
<td>54</td>
<td>M</td>
<td>Routine medical examination</td>
<td>5</td>
<td>17</td>
<td>2.5</td>
<td>Normal</td>
</tr>
<tr>
<td>9</td>
<td>59</td>
<td>F</td>
<td>Palpitations</td>
<td>6</td>
<td>24</td>
<td>3.2</td>
<td>Poor views, normal</td>
</tr>
<tr>
<td>10</td>
<td>52</td>
<td>m</td>
<td>Routine medical examination</td>
<td>11</td>
<td>17</td>
<td>1.9</td>
<td>Normal</td>
</tr>
</tbody>
</table>

F, female; IHD, ischaemic heart disease; M, male.

Figure 1  (A) Left ventricular outflow tract and (B) four chamber cardiovascular magnetic resonance (CMR) views of a patient with apical hypertrophic cardiomyopathy (HCM). Accurate slice alignment shows the true apical cavity (arrows).

Figure 2  A typical patient with apical HCM. There is apical regional hypertrophy, which in systole produces a classic spade shaped ventricular cavity. In the short axis views, the apical cavity is obliterated during systole.
level. Measurements were taken from end diastolic short axis views. Four chamber and two chamber images were also assessed for the presence of apical cavity obliteration and a spade like systolic cavity configuration (fig 2).

RESULTS

Apical HCM was found in all 10 patients with the use of CMR. Apical hypertrophy (table 1, fig 3) ranged from mild to severe (maximum wall thickness 28 mm). No relation was seen between the degree of hypertrophy and echocardiographic findings or the depth of T wave inversion on the ECG (fig 4).

DISCUSSION

We describe 10 patients with an abnormal ECG and non-diagnostic echocardiography where CMR detected apical HCM. The implications are that conventional clinical assessment algorithms are missing patients with apical hypertrophy and that routine echocardiography cannot exclude apical HCM when assessing the patient with an abnormal ECG.

Apical HCM may be underreported in the west. It was first described in Japan in 1976, where it is considered a more benign HCM variant affecting 25% of patients with HCM.11–13 In western countries, although the reported incidence is lower at only 1–2%, the condition appears to be less benign than in Japan and may follow a similar course to other forms of HCM.1 Therefore, western patients with apical HCM are thought to require the same risk stratification and family screening as other HCM patients and the implications of a missed diagnosis are significant.11–14

Echocardiography has been the first line imaging modality for patients with suspected HCM but it is known that the apex may be difficult to image.15 A careful, systematic approach is necessary with sequential short axis examination of the left ventricle in addition to apical views. Subtle abnormalities of diastolic function may lead the operator to suspect apical HCM.10 15–17 Echocardiographic studies with transpulmonary intravenous contrast agents suggest that, in the majority of these cases, poor apical endocardial definition leads to false negative reports (fig 4).18–20 CMR can image in any plane10 21–23 and the introduction of steady state free precession sequences has resulted in a substantial improvement in blood to myocardium contrast, which is ideal for consistent and accurate imaging of the apex.24–26 This has resulted in greater resolution and appreciation of cardiac morphology and, when combined with precise three dimensional piloting, CMR reliably determines the structure of the myocardial apex.27

Experience with CMR in apical hypertrophy and apical HCM is growing. For apical morphology in HCM, Japanese
studies have used CMR. CMR has been compared with echocardiography for the detection of hypertrophied segments and was found to be superior to echocardiography in detecting apical segment hypertrophy. In a recent large series of apical HCM, nine of 105 patients were initially reported to have negative echocardiograms, which on review after CMR were positive. This experience is complemented by several case reports of apical HCM missed with echocardiography or of apical underevaluation. To our knowledge, our series is unique in describing a significant experience of apical HCM missed by echocardiography.

In this series, all the patients had ECG abnormalities in the form of T wave inversion but only half the patients had giant negative T waves. Patients with subtle repolarisation changes may have severe morphological apical HCM, highlighting the importance of additional imaging of such patients where echocardiography is normal. Other causes of extensive T wave inversion may need to be excluded, such as coronary disease, particularly of the proximal left anterior descending artery. T wave inversion can be present in other conditions such as Brugada syndrome, arrhythmogenic right ventricular cardiomyopathy, subarachnoid haemorrhage, and metabolic abnormalities including hypocalcaemia. More localised T wave inversion may be present in other conditions such as left ventricular hypertrophy.

The consequences of missed apical HCM for a patient include unnecessary investigation and missed treatment. On a population scale, however, missed apical HCM implies that the prevalence of apical HCM may have been underreported. If this is the case, then previous echocardiography based studies describing the natural history and risk of apical HCM in the west may be biased, underestimating the prevalence but possibly overestimating the risks associated with the diagnosis.

**Conclusion**

In patients with ECG repolarisation abnormalities without an explained cause, routine echocardiography without contrast may not exclude apical HCM. Definitive exclusion of this important diagnosis requires further imaging such as CMR or contrast echocardiography.

**ACKNOWLEDGEMENTS**

Dr McKenna (chair holder) and Dr James Moon (Junior Research Fellowship) are supported by the British Heart Foundation. The Centre for Advanced Magnetic Resonance in Cardiology (CAMRIC) was supported by a cardiovascular initiative grant from the British Heart Foundation. Support was also received from CORDA The Heart Charity.

The authors thank Dr D G Gibson for advice on the preparation of the manuscript.

**Authors’ affiliations**

J C C Moon, N G Fisher, D J Pennell, Centre for Advanced Magnetic Resonance in Cardiology (CAMRIC), Royal Brompton Hospital, London, UK

W J McKenna, Department of Cardiological Sciences, St George’s Hospital Medical School, London, UK

**REFERENCES**


**Figure 4** ECGs (panels A, B), four chamber echocardiograms (panels C, D), and CMR views (panels E, F) from two patients with apical HCM. Both patients have T wave inversion, with giant negative T waves (B). The echocardiographic images do not identify the apical endocardial border, which is clear by CMR.
Apical hypertrophic cardiomyopathy


8 Ibrahim T, Schwaiger M. Diagnosis of apical hypertrophic cardiomyopathy using magnetic resonance imaging. Heart 2000;83:E1.


FROM BMJ JOURNALS

Myocardial ischaemia in children with sickle cell disease

M de Montalembert, C Maunoury, P Acar, V Brousse, D Sidi, G Lenoir

Background: The heart may be involved in children affected with sickle cell disease (SCD) via several mechanisms. Principally, chronic anaemia increases cardiac output and may cause left ventricular enlargement and cardiac insufficiency.

Aims: To investigate whether the heart also suffers from ischaemia in SCD, as has already been shown for other organs (bone, brain, etc), and to look for risk factors predisposing to this complication.

Methods: Twenty two children with SCD, and chest pain or ECG or echocardiographic signs (left ventricle dilation or hypokinesis) suggesting myocardial ischaemia were subjected to thallium-201 (201Tl) single photon emission computed tomography (SPECT).

Results: Eight children had a normal SPECT, 14 an abnormal one. Myocardial perfusion defects were reversible in nine, fixed in five. Patients with perfusion defects tended to be older and have more severe disease. Five had had cardiac symptoms (episodes of cardiac failure in three, ventricular fibrillation in one, angina in one). Myocardial perfusion was reassessed after six months of hydroxyurea treatment in three patients, and was found to be improved.

Conclusions: Myocardial perfusion defects are present in children with SCD and may be demonstrated using SPECT. Hydroxyurea improved perfusion in three patients.

Please visit the Heart website (www.heartjnl.com) for a link to the full text of this article.
Detection of apical hypertrophic cardiomyopathy by cardiovascular magnetic resonance in patients with non-diagnostic echocardiography

J C C Moon, N G Fisher, W J McKenna and D J Pennell

*Heart* 2004 90: 645-649
doi: 10.1136/hrt.2003.014969

Updated information and services can be found at: [http://heart.bmj.com/content/90/6/645](http://heart.bmj.com/content/90/6/645)

These include:

**References**
This article cites 24 articles, 2 of which you can access for free at: [http://heart.bmj.com/content/90/6/645#BIBL](http://heart.bmj.com/content/90/6/645#BIBL)

**Email alerting service**
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

**Topic Collections**
Articles on similar topics can be found in the following collections

- Clinical diagnostic tests (4779)
- Echocardiography (2127)
- Hypertrophic cardiomyopathy (314)
- Drugs: cardiovascular system (8842)

**Notes**

To request permissions go to: [http://group.bmj.com/group/rights-licensing/permissions](http://group.bmj.com/group/rights-licensing/permissions)

To order reprints go to: [http://journals.bmj.com/cgi/reprintform](http://journals.bmj.com/cgi/reprintform)

To subscribe to BMJ go to: [http://group.bmj.com/subscribe/](http://group.bmj.com/subscribe/)