GUIDELINES AND RECOMMENDATIONS

A systematic approach to echocardiography in hypertrophic cardiomyopathy: a guideline protocol from the British Society of Echocardiography

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Abstract

Hypertrophic cardiomyopathy (HCM) is a relatively common inherited cardiac condition with a prevalence of approximately one in 500. It results in otherwise unexplained hypertrophy of the myocardium and predisposes the patient to a variety of disease-related complications including sudden cardiac death. Echocardiography is of vital importance in the diagnosis, assessment and follow-up of patients with known or suspected HCM. The British Society of Echocardiography (BSE) has previously published a minimum dataset for transthoracic echocardiography, providing the core parameters necessary when performing a standard echocardiographic study. However, for patients with known or suspected HCM, additional views and measurements are necessary. These additional views allow more subtle abnormalities to be detected or may provide important information in order to identify patients with an adverse prognosis. The aim of this Guideline is to outline the additional images and measurements that should be obtained when performing a study on a patient with known or suspected HCM.

Key Words
- hypertrophic cardiomyopathy
- transthoracic echocardiography
- 2D echocardiography
- guidelines

Introduction

1.1 The British Society of Echocardiography (BSE) Education Committee has previously published a minimum dataset for a standard adult transthoracic echocardiogram (1). This Guideline specifically states that the minimum dataset is usually sufficient only when the echocardiographic study is entirely normal. The aim of the BSE Education Committee is to publish a series of appendices to cover specific pathologies supporting this minimum dataset.
1.2 The intended benefits of such supplementary recommendations are to:

- Support cardiologists and echocardiographers to develop local protocols and quality control programs for adult transthoracic study.
- Promote quality by defining a set of descriptive terms and measurements, in conjunction with a systematic approach to performing and reporting a study in specific disease states.
- Facilitate the accurate comparison of serial echocardiograms performed in patients at the same or different sites.

1.3 This Guideline gives recommendations for the image and analysis dataset required in patients either being assessed for, or with a known diagnosis of hypertrophic cardiomyopathy (HCM). The views and measurements are supplementary to those outlined in the minimum dataset and are given assuming that a full study will be performed in all patients.

1.4 When the condition or acoustic windows of the patient prevent the acquisition of one or more components of the supplementary dataset, or when measurements result in misleading information (e.g. off-axis measurements), this should be stated.

1.5 This document is a guideline for echocardiography in HCM and will be updated in accordance with changes directed by publications or changes in practice (Table 1).
Table 1  Additional views and measurements to be obtained in patients with known or suspected hypertrophic cardiomyopathy.

<table>
<thead>
<tr>
<th>View (modality)</th>
<th>Measurement</th>
<th>Explanatory note</th>
<th>Image</th>
</tr>
</thead>
<tbody>
<tr>
<td>PLAX (2D/MM)</td>
<td>IVSd</td>
<td>IVSd measure &gt; 3 cm is a key marker of increased risk (2) Demonstrate if ASH is present Measure RV wall thickness if on axis</td>
<td><img src="image1.png" alt="Image" /></td>
</tr>
<tr>
<td>PLAX (2D/MM)</td>
<td>LA size</td>
<td>Measure LA size (anterior–posterior diameter). LA diameter is one of the criteria used to estimate risk of sudden cardiac death (3)</td>
<td><img src="image2.png" alt="Image" /></td>
</tr>
<tr>
<td>PLAX (MM and CFM)</td>
<td>MV leaflet tips and AV leaflet tips</td>
<td>Demonstrate if SAM is present on M-Mode and for colour flow turbulence within the LVOT Demonstrate if early closure of the AV</td>
<td><img src="image3.png" alt="Image" /></td>
</tr>
<tr>
<td>PSAX MV (2D)</td>
<td>Frozen 2D image: obtain wall thickness measurements from level of the basal LV. Measure at four points, using clock face references (12, 3, 6, 9 o’clock)</td>
<td>To assess for asymmetric and symmetric segmental LV hypertrophy Segmental hypertrophy &gt; 1.5 cm (2) with normal or small LV internal cavity dimensions is strongly suggestive of HCM (in absence of other pathologies such as hypertension)</td>
<td><img src="image4.png" alt="Image" /></td>
</tr>
<tr>
<td>PSAX PM (2D)</td>
<td>2D frozen image at the mid-LV level. Measure at four points, using clock face references (12, 3, 6, 9 o’clock)</td>
<td>Avoid off-axis measurements, papillary muscle and trabeculations</td>
<td><img src="image5.png" alt="Image" /></td>
</tr>
<tr>
<td>PSAX Apex (2D)</td>
<td>Apical-level measure at two points (12 and 6 o’clock)</td>
<td>Apical hypertrophy may be present if apical/basal lateral ratio is &gt;1.5. Consideration should be given to use of LV opacification contrast</td>
<td><img src="image6.png" alt="Image" /></td>
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<tr>
<td>Modified PSAX (2D and PW/CW)</td>
<td>RV wall thickness and RVOT forward flow velocities</td>
<td>Modify both the RV inflow and outflow to assess for RVH and RVOT obstruction. RVH present if &gt; 0.5 cm</td>
<td></td>
</tr>
<tr>
<td>Modified A4C (2D)</td>
<td>RV wall thickness</td>
<td>If clear images can be obtained, measure RV wall thickness. Otherwise measurement from PLAX and subcostal views is preferred. RVH present if &gt; 0.5 cm</td>
<td></td>
</tr>
<tr>
<td>A4C and A2C (2D)</td>
<td>LA volume</td>
<td>Index LA volume to BSA (4)</td>
<td></td>
</tr>
<tr>
<td>A4C (CFM)</td>
<td>Aetiology and severity of mitral regurgitation</td>
<td>If SAM is present, MR may be eccentric and is usually mid/late systolic</td>
<td></td>
</tr>
<tr>
<td>A4C (PW TDI)</td>
<td>Systolic (s’), early (e’) and atrial (a’) relaxation velocities at anterolateral LV annulus</td>
<td>Reduction in s’ or e’ velocities below normal range for age and sex (5)</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Assess for elevated LVEDp by measuring E/e’. Average septal and lateral velocities for e’. Abnormal if &gt; 10 (4)</td>
<td></td>
</tr>
<tr>
<td>A4C (PW TDI)</td>
<td>Systolic (s’), early (e’) and atrial (a’) relaxation velocities at inferoseptal LV annulus</td>
<td>Reduction in Sa or Ea velocities below normal range for age and sex (5)</td>
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<td>A5C and A3C (CFM)</td>
<td>Locate turbulent flow both within the LV cavity and the LVOT</td>
<td>Sample PW Doppler throughout the LV cavity, paying particular attention to areas with turbulent flow. HPRF/CW Doppler may be appropriate if aliasing occurs. Take care not to include MR jet in sample volume. A Valsalva manoeuvre should be performed in the sitting and semi-supine position (and then on standing if no gradient is produced) to assess dynamic LVOT gradients. The peak gradient (rest or Valsalva) should be recorded. In addition, exercise stress echocardiography should be considered in patients with LVOT gradients &lt; 50 mmHg at rest (with or without Valsalva) (3)</td>
<td></td>
</tr>
<tr>
<td>A5C and A3C (PW/CW)</td>
<td>Quantify LVOT/LV intracavity dynamic flow gradient</td>
<td>Sample PW Doppler throughout the LV cavity, paying particular attention to areas with turbulent flow. HPRF/CW Doppler may be appropriate if aliasing occurs. Take care not to include MR jet in sample volume. A Valsalva manoeuvre should be performed in the sitting and semi-supine position (and then on standing if no gradient is produced) to assess dynamic LVOT gradients. The peak gradient (rest or Valsalva) should be recorded. In addition, exercise stress echocardiography should be considered in patients with LVOT gradients &lt; 50 mmHg at rest (with or without Valsalva) (3)</td>
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<td>A2C (PW TDI)</td>
<td>Systolic (s'), early (e') and atrial (a') relaxation velocities at inferior LV annulus</td>
<td>Reduction in s' or e' velocities below normal range for age and sex (5)</td>
<td></td>
</tr>
<tr>
<td>A2C (PW TDI)</td>
<td>Systolic (s'), early (e') and atrial (a') relaxation velocities at anterior LV annulus</td>
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</table>
Abbreviations

Views
A2C  Apical two chamber
A4C  Apical four chamber
A5C  Apical five chamber
A3C  Apical three chamber or apical long axis
PLAX Parasternal long axis
PSAX Parasternal short axis
SC  Subcostal
SSN  Suprasternal

Modality
CFM  Colour flow Doppler
CW  Continuous-wave Doppler
PW  Pulse wave Doppler
TDI  Tissue Doppler imaging

Measurement and explanatory text
a'  Lateral and/or septal late annular relaxation velocity
Ao  Aorta
ASH  Asymmetrical septal hypertrophy
AV  Aortic valve
BSA  Body surface area
DT  Deceleration time
e'  Lateral and/or septal early annular relaxation velocity
HCM  Hypertrophic cardiomyopathy
HPRF  High pulse repetition frequency
IVC  Inferior vena cava
IVSd  Interventricular septal width in diastole
LA  Left atrium
LLPV  Left lower pulmonary vein
LPA  Left pulmonary artery
LUPV  Left upper pulmonary vein
LV  Left ventricle
LVEDp  Left ventricular end-diastolic pressure
LVIDd/s  Left ventricular internal dimension in diastole and systole
LVOT  Left ventricular outflow tract
LVPWd  Left ventricular posterior wall width in diastole
MAPSE  Mitral annular plane systolic excursion
MR  Mitral regurgitation
MV  Mitral valve
PA  Pulmonary artery
PAP  Pulmonary artery pressure
PHT  Pressure half-time
PR  Pulmonary regurgitation
PS  Pulmonary stenosis
PV  Pulmonary valve
RA  Right atrium
RLPV  Right lower pulmonary vein
RV  Right ventricle
RVP  Right upper pulmonary vein
RVH  Right ventricular hypertrophy
RVIDd  Right ventricular cavity diameter in diastole
RWMA  Regional wall motion abnormality
RVOT  Right ventricular outflow tract
RVOTd  Right ventricular outflow tract dimension
s'  Lateral and/or septal systolic annular velocity
SAM  Systolic anterior motion
STJ  Sinotubular junction
SVol  Stroke volume
TAPSE  Tricuspid annular plane systolic excursion
TR  Tricuspid regurgitation
TV  Tricuspid valve
Vmax  Maximum velocity
VSD  Ventricular septal defect
VTI  Velocity time integral
Declaration of interest
This manuscript was prepared by the British Society of Echocardiography Education Committee. The authors declare that there is no conflict of interest that could be perceived as prejudicing the impartiality of this guideline.

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References

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