NHS SCOTLAND

NICCS
NETWORK FOR INHERITED CARDIAC CONDITIONS SCOTLAND

Primary care referral pathway for individuals and families with hypertrophic cardiomyopathy (HCM)

NOTE

This guidance has been prepared by NHS National Services Scotland (NSS) National Networks. Accountable to Scottish Government, NSS works at the heart of the health service providing national strategic services to the rest of NHSScotland and other public sector organisations to help them deliver their services more efficiently and effectively. Working across professional and organisational boundaries, National Networks support the delivery of safe, effective healthcare that's designed around patients, carers and families.

This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined based on all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to guideline recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guideline or any local guidelines derived from it should be fully documented in the patient's case notes at the time the relevant decision is taken.

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Background

NICCS have developed referral and management pathways for individuals and families with inherited arrhythmias and inherited cardiomyopathies. The purpose of this document is to aid primary care health professionals working in NHS Scotland in supporting patients who may have Hypertrophic cardiomyopathy (HCM).

This document was developed by the Inherited Cardiomyopathy Short Life Working Group on behalf of the NICCS Steering Group. Full membership of the group is available in Appendix 1.

Further guidance on the diagnosis and management of inherited cardiomyopathies can be found here: 2023 ESC guidelines for the management of cardiomyopathies

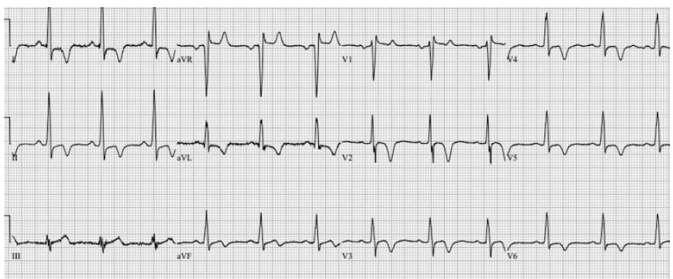
Introduction

Hypertrophic cardiomyopathy (HCM) is a cardiomyopathy affecting all genders and ethnicities with a global prevalence of 1:200-1:500. The genetics of HCM are associated with heterogeneity, incomplete penetrance, variable expression and phenocopies. A diagnosis of HCM is made by finding left ventricular hypertrophy (LVH) in the absence of predisposing cardiovascular conditions. HCM is a treatable condition compatible with normal life expectancy. It may present clinically as:

- Symptoms of chest pain, palpitations, syncope or pre-syncope.
- Asymptomatic murmur
- Abnormal ECG
- Sudden cardiac death or resuscitated cardiac arrest

12-lead ECG and echocardiogram

In most cases, HCM is suspected due to an abnormality in the ECG and/or echocardiogram. The echocardiogram may report left or right ventricular hypertrophy, usually with a normal or high ejection fraction. There may be outflow tract obstruction, but this is not always present. Typical ECG changes seen in left ventricular hypertrophy include high voltage QRS complexes with repolarisation abnormalities. A characteristic pattern, shown below, may be seen in the apical form of HCM.



Establish family history

E.g. family history of HCM, including adverse HCM events (heart failure, cardiac arrest or sudden unexplained death).

Refer for evaluation

If HCM is suspected, the patient should be referred to their local general cardiology team for initial work-up. Referral directly to ICC may be appropriate if they or a family member are already under evaluation by or known to the ICC team.

For patients with a family history of HCM, please include name and DOB for the affected relative in referral.

FOR CHILDREN WITH SUSPECTED HCM, REFER TO PAEDIATRIC CARDIOLOGY

Coding

Guidance is available at www.nn.nhs.scot/niccs/healthcare-professionals/guidance/niccs-guidance/Inclusion of ICD coding facilitates audit and research.

Diagnosis	ICD-10 Code	READ Code
Hypertrophic non-obstructive cardiomyopathy	I42.2	G5543
Hypertrophic obstructive cardiomyopathy	I42.1	G551
Ventricular hypertrophy	I51.7	G5y34

Clinical practice guidelines

2023 Scottish Genomic Test Directory
Scottish Rare and Inherited Disease Test Directory - Scottish Strategic Network for Genomic Medicine (nhs.scot)

2014 European Society of Cardiology Guidelines
ESC Guidelines on Hypertrophic Cardiomyopathy (escardio.org)

2020 American Heart Association / American College of Cardiology
2020 AHA/ACC Guideline for the Diagnosis and Treatment of Patients With Hypertrophic Cardiomyopathy: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines | Journal of the American College of Cardiology (jacc.org)

2023 European Society of Cardiology
ESC Guidelines for the Management of Cardiomyopathies

Appendix 1

Membership of Inherited Cardiomyopathy Short Life Working Group:

Name	Designation	Organisation
Susanne Christie	ICC/Arrhythmia Nurse Specialist	NHS Tayside
Dr Caroline Coats	Consultant Cardiologist	NHS GG&C
Prof Martin Denvir	Consultant Cardiologist	NHS Lothian
Dr Richard Ferguson	Paediatric Cardiologist and Electrophysiologist	NHS Lothian
Dr Fraser Goldie	Cardiology SpR	NHS GG&C
Dr Maria Ilina	Consultant Paediatric Cardiologist	NHS GG&C
Dr Alice Jackson	Cardiology SpR	NHS GG&C
Annie Johnes	Registered Genetic Counsellor	NHS Grampian
Dr Matthew Lee	Cardiology SpR	NHS GJNH
Adele Lewis	ICC Specialist Nurse	NHS Grampian
Debbie Mackin	Genetic Counsellor	NHS Lothian
Ruth McGowan	Consultant in Clinical Genetics	NHS GG&C
Dr Karen McLeod	Consultant Paediatric Cardiologist	NHS GG&C
Robbie Panton	Patient Representative	N/A
Dr Esther Youd	Consultant Pathologist	NHS GG&C