NHS SCOTLAND

NICCS
NETWORK FOR INHERITED CARDIAC CONDITIONS SCOTLAND

Primary care referral pathway for individuals and families with dilated cardiomyopathy (DCM)

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 Dilated cardiomyopathy (DCM).

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

Dilated cardiomyopathy (DCM) is characterised by left ventricular dilatation and a reduction in left ventricular ejection fraction (LVEF), unexplained solely by abnormal loading conditions (e.g. hypertension, valve disease) or coronary artery disease. The aetiology of DCM is heterogenous and includes genetic and acquired causes. Around 30-40% of cases can be attributed to rare pathogenic gene variant. Polygenics and disease modifiers, such as pregnancy or alcohol excess, are often relevant and the identification of an acquired cause does not exclude the possibility of a co-existing gene variant. It may present clinically as:

- Heart failure
- Palpitations or documented arrhythmia
- Myocarditis
- Sudden cardiac death or resuscitated cardiac arrest

12-lead ECG and echocardiogram

In most cases, DCM is suspected due to an abnormality in the ECG and echocardiogram. The echocardiogram may report a dilated left ventricle with a reduction in left ventricular systolic function.

Establish family history

E.g. family history of DCM (including peripartum cardiomyopathy), myocarditis, heart failure, implantation of a cardiac device, cardiac transplantation or sudden death.

Refer for evaluation

A patient with new left ventricular systolic dysfunction 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 DCM, please include name and DOB for the affected relative in referral.

FOR CHILDREN WITH SUSPECTED DCM, 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
Dilated cardiomyopathy	I42.0	G5544

Clinical practice guidelines

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

2023 European Society of Cardiology
Guidelines for the Management of Cardiomyopathies

2020 European Society of Cardiology
Guidelines for the Diagnosis and Treatment of Acute and Chronic Heart Failure

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