3-Tesla cardiac magnetic resonance imaging in primary dilated cardiomyopathy

T Kalekar, MBBS, MD (Radiodiagnosis); **A Gupta,** MBBS, MD (Radiodiagnosis); **M Kumar,** MBBS, MD (Radiodiagnosis)

Department of Radiodiagnosis, Dr. D. Y. Patil Medical College, Hospital and Research Centre, Pune, Maharashtra, India

Corresponding author: *A Gupta (arunimag462@gmail.com)*

Background. Cardiac magnetic resonance imaging (CMR) is an excellent non-invasive imaging tool in the assessment of patients with dilated cardiomyopathy (DCM). Few studies have analysed the findings in primary (idiopathic) DCM.
Objectives. To study the CMR features in primary DCM.
Methods. We conducted a descriptive observational study on 20 adult patients with suspected or confirmed primary DCM. Each patient underwent a dedicated 3-Tesla CMR scan, and the findings were evaluated.
Results. Seventeen patients had systolic dysfunction with a reduced ejection fraction and elevated end-diastolic volume, 19 patients had contractile dysfunction in the form of global left ventricular hypokinesia, 13 patients showed no abnormal delayed contrast enhancement with gadolinium administration, and 7 patients showed abnormal late gadolinium enhancement patterns.
Conclusion. In patients with primary DCM, CMR is a powerful diagnostic tool that can definitively establish the diagnosis, assess the severity of the disease, predict the risk of future adverse cardiovascular outcomes, check for complications, and assist in future follow-ups.
Keywords. Cardiac magnetic resonance imaging, CMR, dilated cardiomyopathy, primary dilated cardiomyopathy.

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Study synopsis
What the study adds. Cardiac magnetic resonance imaging (CMR) is an excellent non-invasive imaging tool in the assessment of patients with primary dilated cardiomyopathy (DCM). Findings include global ventricular enlargement, systolic dysfunction (ejection fraction <40%), and elevated end-diastolic (≥140 mL) and end-systolic volumes. Global abnormal wall contractility is often seen. In DCM there is either no abnormal gadolinium enhancement or curvilinear mid-myocardial or subepicardial late gadolinium enhancement, unrelated to a coronary artery distribution.
Implications of the findings. In patients with primary DCM, CMR provides powerful diagnostic and prognostic information. Enhanced awareness and understanding of this relatively uncommon condition among clinicians and radiologists would be of benefit in patient management and treatment.

Primary dilated cardiomyopathy (DCM) is a myocardial disease characterised by left ventricular (LV) dilation and systolic dysfunction in the absence of coronary artery disease or other causes of LV overload.[1] Patients present clinically with symptoms of heart failure. Complications include thromboembolism, conduction disturbances, arrhythmias, or even sudden death.[2] Identifying potential complications and diagnosing DCM early is crucial, as it can lead to better clinical results and decrease medical expenses. Cardiac magnetic resonance imaging (CMR) is a valuable imaging technique for the diagnosis and follow-up of DCM, as it can provide comprehensive information on myocardial morphology, function, perfusion and fibrosis.

The aim of this study was to examine the CMR features in 20 adult patients with primary DCM.

Methods

We conducted a descriptive observational study on 20 adult patients with suspected or confirmed primary DCM at Dr. D. Y. Patil Medical College, Hospital and Research Centre in Pune, India, from September 2020 to June 2022. Adult patients referred from the cardiology department with either clinical suspicion or a diagnosis of primary DCM (based on clinical data and two-dimensional transthoracic echocardiography) were included in the study. Patients with known risk factors for secondary DCM, including alcohol abuse, ischaemic heart disease, myocardial infarction, hypertension and myocarditis, were excluded from the study. Patients with metabolic, endocrine or haematological disorders, collagen vascular diseases, or exposure to drugs such as anthracycline or cyclophosphamide were also excluded.

A detailed medical history was taken beforehand, and patients with magnetic resonance imaging-incompatible metallic implants were not included in the study. Haemodynamically unstable, claustrophobic or unco-operative patients, and patients with chronic kidney disease with a severely decreased estimated glomerular filtration rate, were also excluded. Before the CMR scan, the patients provided written informed consent. Each patient received a brief explanation of the

scan method before it was performed. Each patient was also coached on proper breath-holding technique.

The CMR scan was conducted using a MAGNETOM Vida MRI 3-Tesla scanner (Siemens, Germany) with retrospective electrocardiogram (ECG) gating.

The patients were placed in a head-first supine position. The ECG leads were connected accordingly. A standard body coil was placed over the chest and secured using straps to prevent artefacts. Cushions were provided for extra comfort. Light music was played to soothe the patients.

Table 1 summarises the sequences used for 3-Tesla CMR in the study.

Results

Twenty patients were evaluated. Thirteen (65%) were male and 7 (35%) were female, and the mean age was 41.5 (range 25 - 60) years. Nineteen of the 20 patients were symptomatic, and 1 patient was asymptomatic. Six patients had mild cardiomegaly, and 14 had moderate cardiomegaly (Figs 1 and 2). Seventeen patients had dilation of the left ventricle only, and 3 patients had dilation of both the left and right ventricles.

Only 3 patients had a preserved LV ejection fraction (LVEF) (>50%). The other 17 patients had reduced systolic function with a reduced LVEF (<40%); 15 of them had a severely reduced LVEF (<30%). These 17 patients also had elevated end-diastolic volumes (EDVs) (≥140 mL). Only 3 patients had a normal EDV (<140 mL). The mean (standard deviation (SD)) LVEF was 30.5 (15.3) mL (range 16.5 - 57.50 mL). The mean LV EDV was 210 (70.8) mL (range 110 - 343 mL).

Sixteen patients also had elevated LV end-systolic volumes (ESVs) (normal range 15 - 64 mL for women and 19 - 88 mL for men). Four patients had normal ESVs. The mean (SD) LV ESV was 149.13 (66.8) mL (range 47.8 - 295 mL).

All but 1 of the patients had contractile dysfunction in the form of global LV hypokinesia. Valvular dysfunction in the form of mitral or tricuspid regurgitation was seen in 17 patients.

Thirteen patients showed no abnormal delayed contrast enhancement with gadolinium administration. Three patients showed a mid-myocardial pattern of late gadolinium enhancement (LGE) (Fig. 1C and D). One patient had both mid-myocardial and subepicardial LGE (Fig. 3). Two patients had patchy subendocardial LGE in a non-territorial distribution. Only 1 patient showed contrast enhancement in the subendocardial location in the diseased coronary artery territory distribution. Fig. 4 summarises the distribution of patterns of LV LGE in the study participants.

Two patients showed mild myocardial oedema on short inversion time inversion recovery. None of the patients had an intracardiac thrombus or clot.

Discussion

Cardiomyopathies are myocardial diseases causing impaired heart function. DCM is characterised by enlargement and dilation of one or both of the ventricles, with associated systolic dysfunction.[3,4] It is the most common form of cardiomyopathy. DCM usually affects adults between 20 and 60 years of age, but it can also occur in the paediatric population and in the elderly. Patients present clinically with symptoms of heart failure. Complications include thromboembolism, conduction disturbances, arrhythmias, or even sudden death.

In primary or idiopathic DCM, a definite cause cannot be identified. It is likely that a number of factors, including genetic alterations[5] and environmental exposures that cause myocardial

Table 1. A summary of sequences used for 3-Tesla cardiac magnetic resonance imaging in the study

1. For morphology
T2 TRUFI single shot
T2 HASTE dark blood
T1 TSE dark blood
T2 TSE dark blood
T2 STIR dark blood long axis
2. For LV function
2-chamber cine TRUFI retro long axis
4-chamber cine TRUFI retro long axis
Cine TRUFI retro long axis
Cine LVOT
Cine RVOT
3. For perfusion
Images are taken in 2-chamber, 4-chamber, and short-axis views for left side and 2-chamber view for right side
DYNAMIC TRUFI SR EPAT
4. For late enhancement imaging
TI SCOUT
Delayed TRUFI high- resolution PSIR

TRUFI = true fast imaging with steady-state free precession; HASTE = half-Fourier single-shot turbo spin-echo; TSE = turbo spin echo; STIR = short inversion time inversion recovery; LVOT = left ventricular outflow tract; RVOT = right ventricular outflow tract; TI SCOUT = inversion time mapping sequence; PSIR = phase-sensitive inversion recovery.

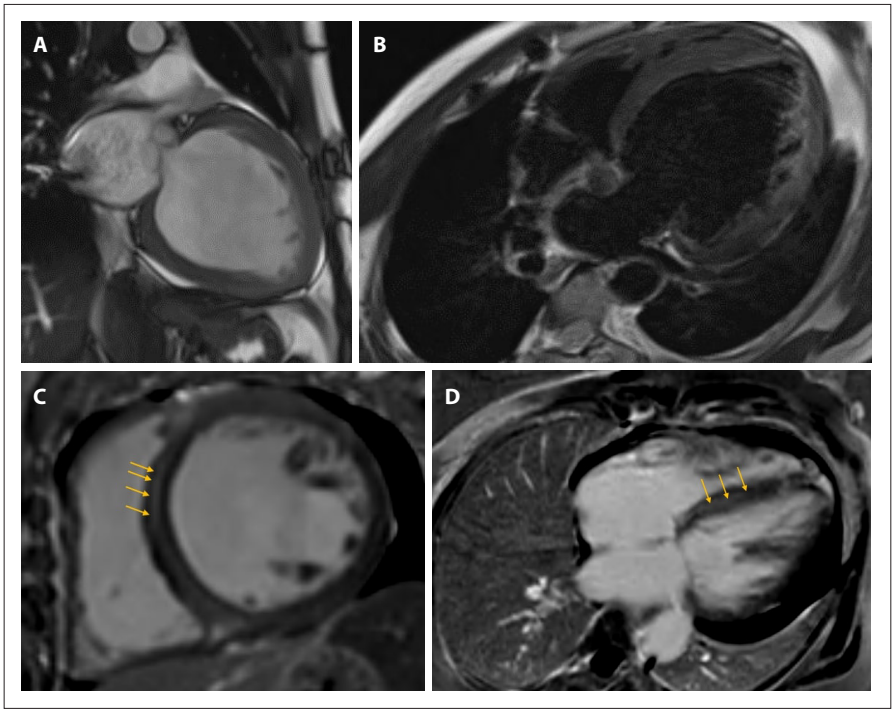

Fig. 1. A 55-year-old woman with primary dilated cardiomyopathy. (A) Two-chamber cine two-dimensional steady-state free precession and (B) morphological T2-weighted black blood four-chamber long-axis images acquired at 3-Tesla show a markedly dilated left ventricle. The calculated left ventricular end-diastolic volume was 256 mL. The patient had severely decreased left ventricular systolic function (left ventricular ejection fraction 19%). (A) also shows mild pericardial effusion. Late-enhancement short-axis (C) and four-chamber long-axis (D) images show abnormal mid-myocardial late gadolinium enhancement in the interventricular septum (arrows).

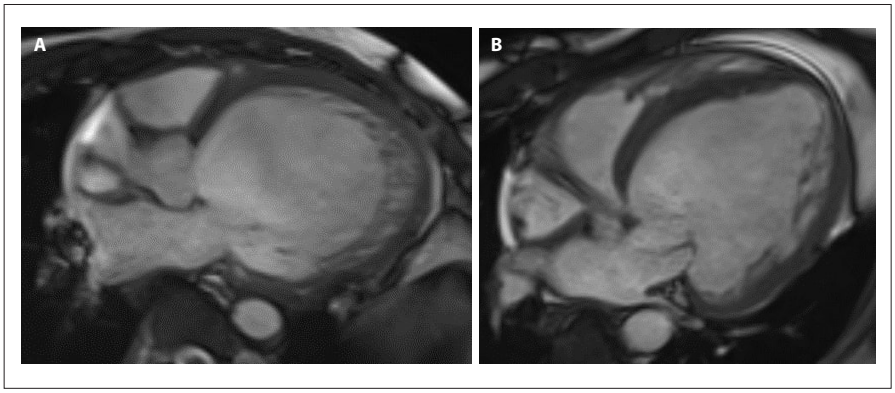

Fig. 2. A 40-year-old man with primary dilated cardiomyopathy. (A) Left ventricular outflow tract steady-state free precession and (B) four-chamber long-axis steady-state free precession images acquired at 3-Tesla show a markedly dilated left ventricle (left ventricular end-diastolic volume 339 mL). The patient was clinically symptomatic and had severely decreased left ventricular systolic function (left ventricular ejection fraction 26.5%). No abnormal delayed enhancement was seen with gadolinium administration.

injury, contribute to these conditions. Secondary DCM occurs as a result of any myocardial insult that causes LV failure and subsequent ventricular enlargement. Secondary causes include alcohol abuse, ischaemic heart disease, hypertension, infectious myocarditis (e.g. viral, parasitic or bacterial), drug-induced myocarditis (e.g. anthracyclines, cyclophosphamide, heavy metals), peripartum cardiomyopathy, metabolic, endocrine or haematological disorders, infiltrative diseases such as sarcoidosis, and collagen vascular diseases.[4,6] A diagnosis of primary or idiopathic DCM is established only after the exclusion of known secondary causes. We included patients with suspected or confirmed DCM in our study only after comprehensive exclusion of secondary causes.

The primary investigations used for DCM evaluation are echocardiography and CMR. Transthoracic echocardiography is generally the initial diagnostic modality employed. It is able to detect chamber dynamics, valvular motion, and gross morphological and functional anomalies. However, several factors, such as the experience of the operator, a smaller field of view and unfavourable patient body characteristics, limit its imaging capability. Transoesophageal echocardiography is not limited by a restricted field of view, but is an invasive technique.[7] Also, neither echocardiographic technique enables tissue characterisation of the myocardium.

CMR is a valuable technique for the diagnosis and prognosis of patients with DCM. It provides high spatial and temporal resolution, enhanced soft-tissue contrast, and improved cardiac tissue characterisation. CMR has therefore been established as an essential non-invasive tool for the comprehensive evaluation of patients with DCM.[8]

Findings at CMR include global enlargement of the LV or both ventricles and systolic dysfunction (decreased ejection fraction <40%). The ventricular enlargement is usually uniform with normal wall thickness. The EDV (≥140 mL) and ESV are elevated.[9,10] In our study, all the patients had variable degrees of cardiomegaly; 85% had systolic dysfunction with a reduced LVEF and elevated LV EDV.

Wall motion abnormalities are also seen in DCM, generally in the form of global hypokinesis. Valvular dysfunction is frequently evident as a result of chamber expansion and annular strain. Ventricular thrombus may be a problem in patients with DCM because of diminished wall mobility.[10] In our study, 95% of the patients had contractile dysfunction. Valvular dysfunction was seen in 85% of the patients. None of the patients had an intracardiac thrombus or clot.

CMR makes it possible to differentiate between ischaemic cardiomyopathy and idiopathic DCM. DCM shows either no abnormal enhancement[11] or curvilinear mid-myocardial or subepicardial LGE, unrelated to

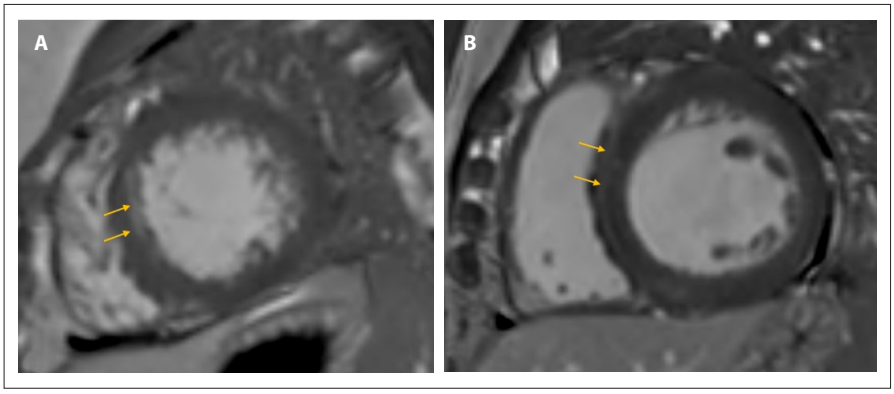

Fig. 3. A 46-year-old man with primary dilated cardiomyopathy. Short-axis delayed contrast images show (A) patchy mid-myocardial late gadolinium enhancement in the interventricular septum (arrows), and (B) patchy subepicardial enhancement of the anteroseptal segment of the basal cavity (arrows).

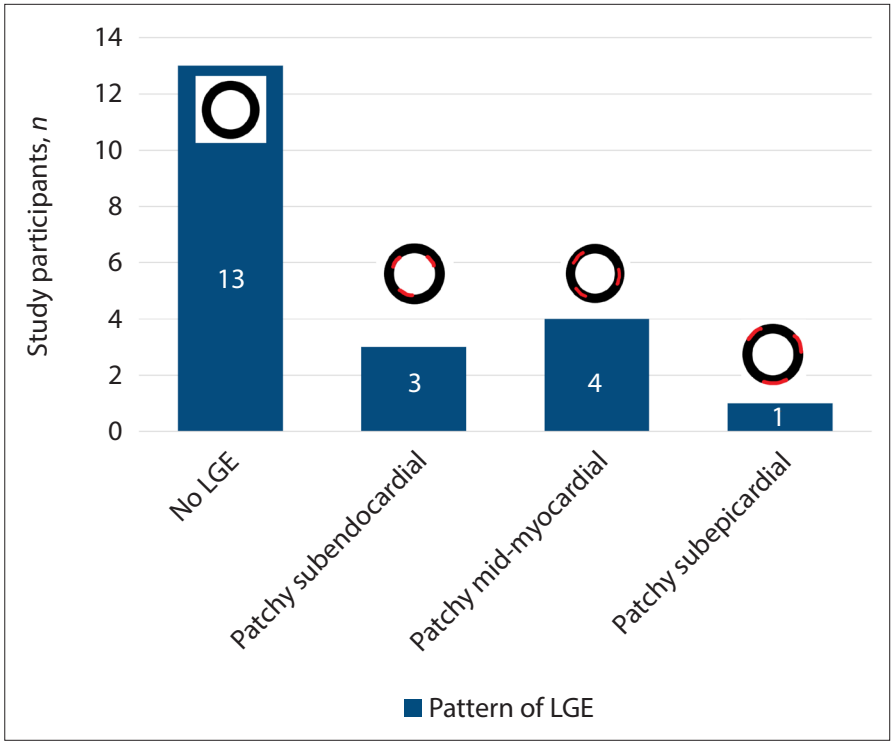

Fig. 4. Distribution of patterns of left ventricular LGE in the study participants (N=20). One patient had both mid-myocardial and subepicardial patterns of LGE. (LGE = late gadolinium enhancement.)

a coronary artery distribution.[12,13] In our study, 13 patients showed no abnormal LGE. Three patients showed a mid-myocardial LGE pattern, indicative of mid-wall fibrosis, and 1 patient showed both mid-myocardial and subepicardial LGE.

In our study, one 35-year-old patient, with no prior history of myocardial infarction, showed contrast enhancement in the subendocardial location in the diseased coronary artery territory distribution. This may be explained by the findings of McCrohon *et al.*,[14] who showed that some patients with DCM may have subendocardial fibrosis characteristic of infarction. These may either represent cases of coronary emboli-induced ischaemic cardiomyopathy, or recanalisation after an occlusive coronary event.[12,14]

One possible application of CMR is to assess the prognosis of patients with DCM. LGE, particularly mid-myocardial, in patients with DCM is linked to an increased likelihood of serious cardiovascular events, including ventricular tachycardia and sudden cardiac death.[11,15] In our study, 7 patients showed abnormal LGE, with 4 of the 7 showing mid-myocardial LGE. The latter finding was indicative of a poorer prognosis.

In summary, CMR has many uses as a diagnostic tool that can significantly benefit patient management. It can help differentiate primary DCM from other causes of LV dysfunction, such as ischaemic cardiomyopathy or restrictive cardiomyopathy. It can also detect areas of fibrosis within the myocardium, which may have prognostic value and guide therapeutic decisions. Furthermore, CMR can assess the degree of ventricular dilation and systolic dysfunction, which are important parameters for monitoring disease progression and response to treatment.

Conclusion

Primary DCM is relatively uncommon, but contributes significantly to cardiovascular morbidity and mortality. An accurate diagnosis is therefore of the utmost importance. At present, the primary diagnosis is based on clinical suspicion and echocardiographic findings. CMR has emerged as a powerful diagnostic tool that, by providing accurate, high-resolution reproducible images, can definitively establish the diagnosis, assess the severity of the disease both qualitatively and quantitatively, predict the risk of future adverse cardiovascular outcomes, check for complications, and assist in future follow-ups.

Declaration. The research for this study was done in partial fulfilment of the requirements for AG's MD (Radiodiagnosis) degree at Dr. D. Y. Patil Medical College, Hospital and Research Centre, Pune.

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