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INNER SPACES
Edited by Dr. Bhavin Jankharia

RNI No.: MAHENG/2006/17782
Regd. No.: MCS/022/2021-23
WPP no. MR/Tech/WPP-174/South/2021-2023

January. 2023 | Vol. 23 | No. 1
Published: 7th of every month | Subscription Price: Rs. 10
Posted at Mumbai Patrika Channel Sorting Office Mumbai 400 001 on 9th of Every Month

Arrhythmogenic Cardiomyopathy

2023 Happy New Year

Arrhythmogenic cardiomyopathies (ACMs) are a group of genetic disorders that affect the RV, LV or both with the development of fibrous or fibrofatty scars that can cause fatal arrhythmias.

ACMs can be only right ventricular (ARVC – arrhythmogenic right ventricular cardiomyopathy), biventricular or only LV predominant.

The last time the diagnostic criteria guidelines were published was in 2010. Since then, with the advances in cardiac MRI and in electrophysiology, new information and data have prompted the publication of new guidelines in 2020, the Padua guidelines, which apart from the usual morpho-functional abnormalities, now include structural abnormalities as part of the major criteria.

The role of cardiac MRI is in defining

1. Morpho-functional criteria – akinesia, dyskinesia and bulging of the RV walls with reduced RV ejection fraction or increased end-diastolic volume (major criteria). If only akinesia, dyskinesia and RV bulging are present without reduced EF or increased EDV, then this is a minor criterion.
2. Structural abnormality – abnormal enhancement that defines the presence of fibrosis is a major criterion.

The diagnosis of fat in the myocardium is a confirmatory sign, but is not part of the guidelines, due to a variety of reasons.

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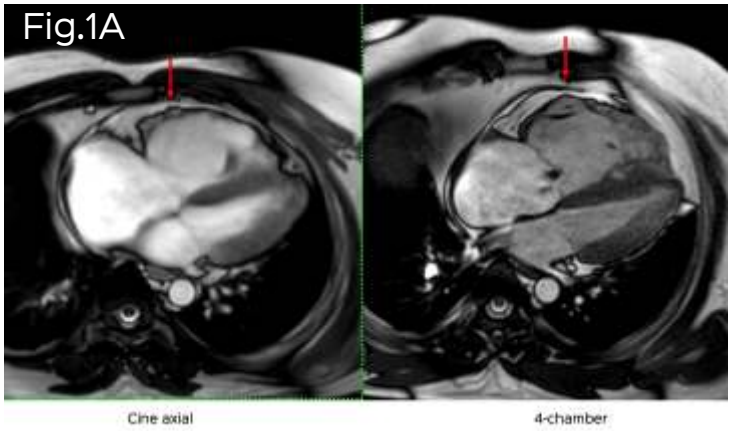
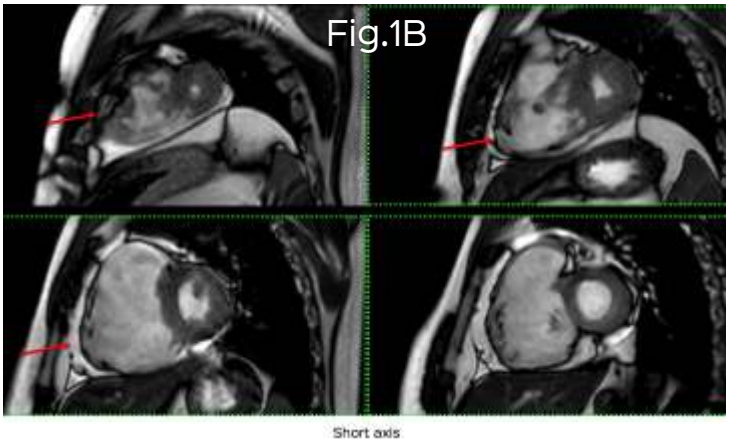
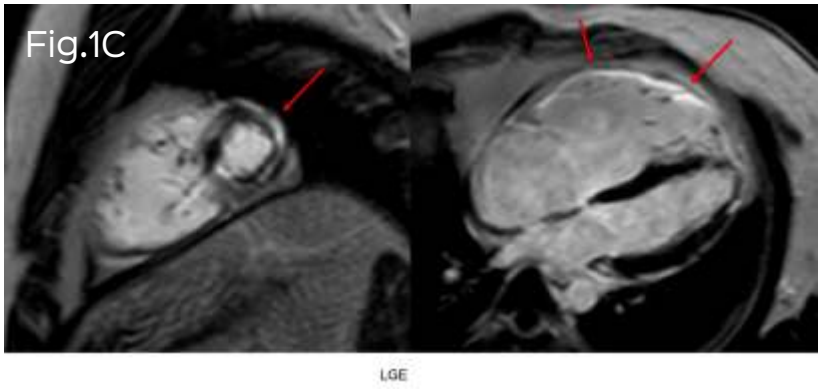
At a glance:

- Arrhythmogenic cardiomyopathies can be RV (ARVC), biventricular or LV (ALVC) predominant
- The new 2020 Padua criteria have made is easier to reach the diagnosis in the correct clinical setting.







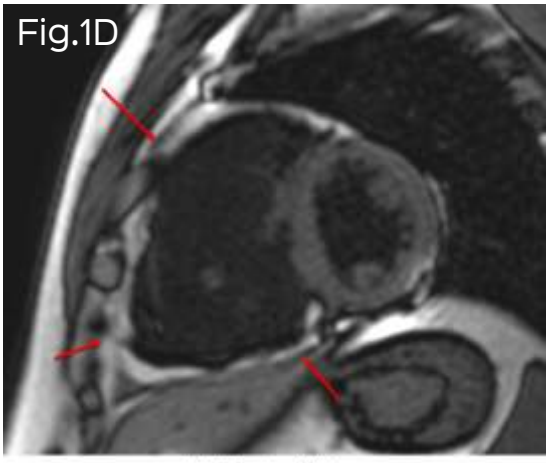


Fig 1 (A-D). Biventricular, RV predominant, arrhythmogenic cardiomyopathy. 40-years old with arrhythmias. Cardiac MRI shows abnormal wall motion in the axial and four-chamber (A) cines and the short axis (B) cines with areas of dyskinesia and pseudobulging (arrows). The late gadolinium enhanced (LGE) images (C) show areas of enhancement involving the RV free wall and the LV apex. The T1 image from a MOLLI sequence (D) shows fat in the RV myocardium.

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Main Clinic
383 | Bhaveshwar Vihar | Sardar V. P. Road | Prarthana Samaj | Charni Road | Mumbai 400 004 | **T:** 022 66173333

Cardiac, Chest & Interventional Twin Beam CT
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Owner, Printer & Publisher: Dr. Bhavin Jankharia
Published at: Dr. Jankharia's Imaging Centre Bhaveshwar Vihar, 383, S.V.P. Road, Prarthana Samaj, Charni Road, Mumbai 400 004.
Printed at: India Printing House, First Floor, 42, G D Ambedkar Marg, Opp. Wadala Post Office, Wadala, Mumbai 400 031