Imaging in Seven Cardiomyopathies: Diagnostic and Prognostic Approaches

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Northwestern Memorial Hospital

No Relationships to Disclose
Cardiomyopathies:

- Echocardiography
- Cardiac magnetic resonance
- Cardiac computed tomography
Echocardiography:

- Versatile
- Portable and available
- Relatively inexpensive
- Quantitative Doppler established and straightforward
- Transesophageal assessment
- Advances in 3D imaging
- Advances in TDI, contrast, speckle tracking, strain imaging
**Echocardiography:**

- Versatile
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Echocardiography:

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- Transesophageal assessment
- Advances in 3D imaging
- Advances in TDI, contrast, speckle tracking, strain imaging
With advances in echocardiography, is there role for multimodality imaging in evaluating myocardial disease?
Differentiation of Ischemic and Dilated Cardiomyopathy

McCrohon et al, Circulation 2003;108:54-59
Cardiovascular Magnetic Resonance, Fibrosis, and Prognosis in Dilated Cardiomyopathy

Ravi G. Assomull, MRCP,*† Sanjay K. Prasad, MD, MRCP,*† Jonathan Lyne, MRCP,* Gillian Smith, MSc,* Elizabeth D. Berman, MSc,* Mohammed Khan, MSc, MPH ‡ Mary N. Sheppard, MD, FRCPATH,‡ Dudley J. Pennell, MD, FRCP, FESC*

London, United Kingdom

J Am Coll Cardiol 2006;48:1977–85

![Graph showing survival (percent) over time (days) with LGE− n=65 and LGE+ n=35, with a p-value of 0.03.]

n=101 patients

p=0.03
Association of Fibrosis With Mortality and Sudden Cardiac Death in Patients With Nonischemic Dilated Cardiomyopathy

Ankur Gulati, MD
Andrew Jabbour, MD, PhD
Tevfik F. Ismail, MD
Kaushik Guha, MD
Jahanzaib Khwaja, BSc
Sadaf Raza, MD
Kishen Morarji, MD
Tristan D. H. Brown, BSc
Nizar A. Ismail, BSc
Marc R. Dweck, MD
Elisa Di Pietro, MD
Michael Roughton, MSc
Ricardo Wage, DCR
Yousef Daryani, MD
Rory O’Hanlon, MD
Mary N. Sheppard, MD
Francisco Alpendurada, MD
Alexander R. Lyon, MD, PhD
Stuart A. Cook, MD
Martin R. Cowie, MD
Ravi G. Assomull, MD
Dudley J. Pennell, MD
Sunjay K. Prasad, MD

Gulati et al, JAMA 2013;309:896-908
Association of Fibrosis With Mortality and Sudden Cardiac Death in Patients With Nonischemic Dilated Cardiomyopathy

Ankur Gulati, MD
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Tevfik F. Ismail, MD
Kashinath Guha, MD
Jahanzaib Khwaja, BSc
Sadaf Raza, MD
Kishen Morarji, MD
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Alexander R. Lyon, MD, PhD
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Martin R. Cowie, MD
Ravi C. Assomull, MD
Dudley J. Pennell, MD
Sanjay K. Prasad, MD

Survival (percent)

Time (years)

n=472 patients

No fibrosis

Fibrosis

p <0.001

Gulati et al, JAMA 2013;309:896-908
Novel Predictors of Left Ventricular Reverse Remodeling in Individuals With Recent-Onset Dilated Cardiomyopathy

Milos Kubanek, MD, PtD,* Marek Sramko, Jiri Weichert, MD, PtD,* Petr Lupinek, MD, Josef Kautzner, MD, PtD*
Prague, Czech Republic

J Am Coll Cardiol 2013;61:54–63
Hypertrophic Cardiomyopathy

Myocardial Scarring in Asymptomatic or Mildly Symptomatic Patients With Hypertrophic Cardiomyopathy

Lubna Choudhury, MD, MRCP, Heiko Mahrholdt, MD, Anja Wagner, MD, Kelly M. Choi, MD, Michael D. Elliott, MD, Francis J. Klocke, MD, MACC, Robert O. Bonow, MD, FACC, Robert M. Judd, PhD, Raymond J. Kim, MD, FACC

Chicago, Illinois

J Am Coll Cardiol

Systole

Diastole

Contrast

Choudhury et al, J Am Coll Cardiol 2002;40:2156-64
Hypertrophic Cardiomyopathy

Myocardial Scarring in Asymptomatic or Mildly Symptomatic Patients With Hypertrophic Cardiomyopathy

Lubna Choudhury, MD, MRCP, Heiko Mahrholdt, MD, Anja Wagner, MD, Kelly M. Choi, MD, Michael D. Elliott, MD, Francis J. Klocke, MD, MACC, Robert O. Bonow, MD, FACC, Robert M. Judd, PhD, Raymond J. Kim, MD, FACC

Choudhury et al, J Am Coll Cardiol 2002;40:2156-64
Prognostic Significance of Myocardial Fibrosis in HCM

N=1293
P=0.008

Chan et al, Circulation 2014;130:484-495
Prognostic Value of LGE-CMR in HCM
A Meta-Analysis

Zhen Weng, PhD,a Jialu Yao, MD,b Raymond H. Chan, MD, MPH,c Jun He, MD,d Xiangjun Yang, MD, PhD,b Yafeng Zhou, MD, PhD,b Yang He, MDb

J Am Coll Cardiog Img 2016;9:1392-1402

Cumulative Analysis of Sudden Cardiac Death

<table>
<thead>
<tr>
<th>Study Name</th>
<th>Cumulative Statistics</th>
<th>Cumulative Odds Ratio (95% CI)</th>
</tr>
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<tr>
<td></td>
<td>Point</td>
<td>Lower Limit</td>
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<tr>
<td>Bruder et al (2010)</td>
<td>5.145</td>
<td>0.646</td>
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<td>Rubinshtein et al (2010)</td>
<td>7.200</td>
<td>1.343</td>
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<tr>
<td>Chan et al (2014)</td>
<td>3.741</td>
<td>1.939</td>
</tr>
<tr>
<td>Hen et al (2014)</td>
<td>3.890</td>
<td>2.051</td>
</tr>
<tr>
<td>Ismail et al (2014)</td>
<td>3.419</td>
<td>1.967</td>
</tr>
<tr>
<td>Pooled</td>
<td>3.419</td>
<td>1.967</td>
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</tbody>
</table>
Indications for CMR

- Inconclusive echo for diagnosis or assessment of distribution of LVH  
  class I
- Define apical HCM  
  class IIa
- Late gadolinium imaging when risk stratification is inconclusive  
  class IIb
Amyloidosis
CMR Imaging With Rapid Visual T1 Assessment Predicts Mortality in Patients Suspected of Cardiac Amyloidosis

James A. White, MD,*†† Han W. Kim, MD,§§ Dipan Shah, MD,# Nowell Fine, MD,* Ki-Young Kim, MD,§ David C. Weng, MD,§ Michele Parker, MS,§§ Manesh Patel, MD,§ Robert M. Judd, PhD,§§ Raymond V. T. Fung, MD,§ London, Ontario, Canada; Durham, NC

J Am Coll Cardiol Img 2014;7:143–56
CMR Imaging With Rapid Visual T1 Assessment Predicts Mortality in Patients Suspected of Cardiac Amyloidosis

James A. White, MD,*†‡ Han W. Kim, MD,*§ Dipan Shah, MD,# Nowell Fine, MD,*
Ki-Young Kim, MD,*§ David C. Wendell, Ph.D,*§ Wael Al-Jaroudi, MD,*§
Michele Parker, MS,*§ Manesh Patel, MD,*§ Femida Gwadry-Sridhar, Ph.D,*‡
Robert M. Judd, Ph.D,*‡ Raymond J. Kim, MD*§§
London, Ontario, Canada; Durham, North Carolina

J Am Coll Cardiol Img 2014;7:143–56

![Survival (%)](image)

- **Diffuse HE Absent**
- **Diffuse HE Present**

**P < 0.0001**
Prognostic Value of Late Gadolinium Enhancement Cardiovascular Magnetic Resonance in Cardiac Amyloidosis

Marianna Fontana, MD; Silvia Pica, MD; Patricia Reant, MD, PhD;
Amna Abdel-Gadir, MBBS; Thomas A. Treibel, MBBS; Sanjay M. Baypersad, MBChB;
Viviana Maestrini, MD; William Barcella, BFIN, MSc; Stefania Rosmini, MD; Heerajnarain Bulluck, MBBS;
Rabya H. Sayed, MBBS; Ketna Patel, MBBS; Shameem Mamhood, MBBChBAO;
Chiara Bucciarelli-Ducci, PhD; Carol J. Whelan, MD; Anna S. Herrey, MD;
Helen J. Lachmann, MSc; Tamar A. Taylor, MSc; Eric B. Schelbert, PhD, MD;
Imaging

Prognostic Value of Late Gadolinium Enhancement Cardiovascular Magnetic Resonance in Cardiac Amyloidosis

Marianna Fontana, MD; Silvia Pica, MD; Patricia Reant, MD, PhD; Amna Abdel-Gadir, MBBS; Thomas A. Treibel, MBBS; Sanjay M. Banyersad, MBChB; Viviana Maestrini, MD; William Barcella, BFIN, MSc; Stefania Rosmini, MD; Heerajnarin Bulluck, MBBS; Rabya H. Sayed, MBBS; Ketsa Patel, MBBS; Shameem Mamooh, MBBCo BA; Chiara Bucairelli-Ducci, PhD; Carol J. Whelan, MD; Anna S. Herrey, MD; Helen J. Lachmann, MD; Ashutosh D. Wechalekar, MD, PhD; Charlotte H. Manisty, PhD;

Fontana et al, Circulation 2015;132:1570-1579
Amyloidosis
Radionuclide Imaging

$^{99m}\text{Tc} - \text{DPD}$

$^{99m}\text{Tc}$ diphosphono-propanodicarboxylic acid
Amyloidosis
Radionuclide Imaging

$^{99m}$Tc - DPD
$^{99m}$Tc diphosphono-propanodicarboxylic acid

$^{99m}$Tc - Pyrophosphate
Multicenter Study of Planar Technetium 99m Pyrophosphate Cardiac Imaging
Predicting Survival for Patients With ATTR Cardiac Amyloidosis

Adam Castano, MD, MS; Muhammad Hao, MD; David L. Narotsky, MD; Jeff Goldsmith, PhD; Richard L. Weinberg, MD, PhD; Rachelle Morgenstern, MPH; Ted Porziakoff, ARRT; Frederick L. Ruberg, MD; Edward J. Miller, MD, PhD; John L. Berk, MD; Angela Dispenzieri, MD; Martha Grogan, MD; Geoffrey Johnson, MD, PhD; Sabahat Bokhari, MD; Matthew S. Maurer, MD

JAMA Cardiol. 2016;1:880-889

**Multicenter Analysis**

<table>
<thead>
<tr>
<th></th>
<th>No. of Patients</th>
</tr>
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<tbody>
<tr>
<td>ATTR⁺</td>
<td>ATTR⁻</td>
</tr>
<tr>
<td>Positive scan</td>
<td>110</td>
</tr>
<tr>
<td>Negative scan</td>
<td>11</td>
</tr>
<tr>
<td>Total</td>
<td>121</td>
</tr>
</tbody>
</table>

Sensitivity = 91%
Specificity = 92%
AUC, 0.960 (95% CI, 0.930-0.981)
Sarcoidosis
CMR Imaging Predicts Death and Other Adverse Events in Suspected Cardiac Sarcoidosis

Simon Greulich, MD,* Claudia Christina Deluigi, MD,† Steffen Gloekler, MD,‡ Andreas Wahl, MD,‡ Christine Zürn, MD,§ Ulrich Kramer, MD,¶ Detlev Nothnagel, MD,¶ Helmut Büttel, MD,# Julia Schumann, MD,* Stefie Grön, MD,* Peter Oep, MD,* Anja Wagner, MD,* Meinrad Gawaz, MD,*
CMR Imaging Predicts Death and Other Adverse Events in Suspected Cardiac Sarcoidosis

Simon Greulich, MD,* Andreas Wahl, MD,‡ Helmut Bütel, MD,§ Anja Wagner, MD,** Meinrad Gawaz, MD,§

J Am Coll Cardiol Img 2013;6:501-511
Complementary Role of CMR to Conventional Screening in the Diagnosis and Prognosis of Cardiac Sarcoidosis

Vasileios Kouranos, MD, MSc, a,b George E. Tzelepi, MD, a,b Konstantina Aggeli, MD, PhD, c Marousa Douskou, PhD, d Petros Sfikakis, MD, PhD, e Athol Wells, MD, PhD, f

J Am Coll Cardiol Img 2017;10:1437-1447

N=321

P<0.001
Hybrid Magnetic Resonance Imaging and Positron Emission Tomography With Fluorodeoxyglucose to Diagnose Active Cardiac Sarcoidosis

Marc R. Dweck, MD, PhD, a,b Ronan Abgral, MD, PhD, a,c Maria Giovanna Trivieri, MD, PhD, a Philip M. Robson, PhD, a Nicolas Karakatsanis, PhD, a Venkatesh Mani, PhD, a Anna Palmisano, MD, a Marc A. Miller, MD, d Anuradha Lala, MD, d Helena L. Chang, MS, e Javier Sanz, MD, d Johanna Contreras, MD, d Jagat Narula, MD, PhD, d Valentín Fuster, MD, PhD, d Maria Padilla, MD, e Zahi A. Fayad, PhD, a Jason G. Kovacic, MD, PhD d

J Am Coll Cardiol Img  http://dx.doi.org/10.1016/j.jcmg.2017.02.021
Hybrid Magnetic Resonance Positron Emission Tomography Fluorodeoxyglucose Cardiac Sarcoidosis

Marc R. Dweck, MD, PhD, Ronan Abgral, MD, PhD, Nicolas Karakatsanis, PhD, Venkatesh Mani, PhD, Aleksej Z. Nikolaev, PhD, Helena L. Chang, MS, Javier Sanz, MD, Johanna O. Cordero, MD, Valentin Fuster, MD, PhD, Maria Padilla, MD, Zaher S. Nasir, MD

Dweck et al, J Am Coll Cardiol Img 2017 [epub ahead of print]
Myocarditis

Kwong R in Braunwald's *Heart Disease* 10th ed, 2011
Myocarditis

N=670
P<0.001

Myocarditis

Hypertrophic cardiomyopathy

Dilated cardiomyopathy

Ischemic cardiomyopathy

Amyloidosis

Sarcoidosis

Myocarditis
Imaging

Equilibrium Contrast Cardiovascular Magnetic Resonance for the Measurement of Diffuse Myocardial Fibrosis
Preliminary Validation in Humans

Andrew S. Flett, MB, BS, BSc; Martin P. Hayward, MBBS, BSc, MS;
Michael T. Ashworth, MD; Michael S. Hansen, PhD; Andrew M. Taylor, MD;
Perry M. Elliott, MB, BS, MD; Christopher McGregor, MD; James C. Moon, MB, BCh, MD

Circulation. 2010;122:138-144
Native T1 Mapping in Differentiation of Normal Myocardium From Diffuse Disease in Hypertrophic and Dilated Cardiomyopathy

Valentina O. Puntnann, MD, PhD, Tobias Voigt, PtD, Zhong Chen, MD, Manuel Mayr, MD, PtD, Rasheed Karim, PtD, Kawal Rhode, PtD, Ana Pastor, MD, Gerald Carr-White, MBBS, PtD, Reza Razavi, MD, Tobias Schaeffter, PtD, Eike Nagel, MD, PtD

London, United Kingdom

J Am Coll Cardiol Img 2013;6:273-84
Native T1 Mapping in Differentiation of Normal Myocardium From Diffuse Disease in Hypertrophic and Dilated Cardiomyopathy

Valentina O. Puntmann, MD, PhD,* Tobias Voigt, PhD,‡ Zhong Chen, MD,* Manuel Mayr, MD, PhD,‡ Rashed Karim, PhD,* Kawal Rhode, PhD,* Ana Pastor, MD,* Gerald Carr-White, MBBS, PhD,‡ Reza Razavi, MD,* Tobias Schaeffter, PhD,* Elke Nagel, MD, PhD*

London, United Kingdom

J Am Coll Cardiol Img 2013;6:273-84
Noncompaction
Noncompaction
Takotsubo Cardiomyopathy
Takotsubo Cardiomyopathy

Brief Report

Differences in the Clinical Profile and Outcomes of Typical and Atypical Takotsubo Syndrome
Data From the International Takotsubo Registry

Jelena R. Ghadri, MD; Victoria L. Cammann; L. Christian Napp, MD; Stjepan Jurisic; Johanna Diekmann; Dana Roxana Batalosu, MD; Burkhart Seifert, PhD; Milosz Jaguszewski, MD; Annahita Sarcon, MD; Catharina A. Neumann; Verena Geyer, MD; Abhiram Prasad, MD; Jeroen J. Bax, MD, PhD; Frank Ruschitzka, MD; Thomas F. Lüscher, MD; Christian Templin, MD, PhD; for the International Takotsubo (InterTAK) Registry

JAMA Cardiol. 2016;1:335-340
Takotsubo Cardiomyopathy

Kohan et al, Caridovasc Diagn Ther 2010;14:138-146
Takotsubo Cardiomyopathy
Usefulness of Electron Beam Computed Tomography Scanning for Distinguishing Ischemic From Nonischemic Cardiomyopathy

MATTHEW J. BUDOFF, MD, DAVID M. SHAVELLE, MD, DANIEL H. LAMONT, MD, H. TINA KIM, BS, PAMELA AKINWALE, JOHN M. KENNEDY, MD, BRUCE H. BRUNDAGE, MD, FACC

Torrance, California

J Am Coll Cardiol 1998;32:1173–8
Usefulness of Electron Beam Computed Tomography Scanning for Distinguishing Ischemic From Nonischemic Cardiomyopathy

MATTHEW J. BUDOFF, MD, DAVID M. SHAVELLE, MD, DANIEL H. LAMONT, MD, H. TINA KIM, BS, PAMELA AKINWALE, JOHN M. KENNEDY, MD, BRUCE H. BRUNDAGE, MD, FACC

Torrance, California

Diagnostic Accuracy of Multidetector Computed Tomography Coronary Angiography in Patients With Dilated Cardiomyopathy

Daniele Andreini, MD, Gianluca Pontone, MD, Mauro Pepi, MD, Giovanni Ballerini, MD, Antonio L. Bartorelli, MD, FACC, Alessandra Magini, MD, Carlo Quaglia, MD, Enrica Nobili, MD, Piergiuseppe Agostoni, MD, PhD

Milan, Italy
Multimodality Imaging: Myocardial Disease

**Cardiac magnetic resonance:**

- HCM: morphology and fibrosis (prognosis)
- DCM: exclude prior MI
- DCM: function and fibrosis (prognosis)
- Sarcoid: diagnosis
- Amyloid: diagnosis, therapeutic options
- Noncompaction: accurate diagnosis
- Takotsubo: diagnosis (…prognosis?)
Multimodality Imaging: Myocardial Disease

**Cardiac magnetic resonance:**
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**CT angiography:**
- DCM: exclude ischemic cardiomyopathy