Why Do I Do it?
Standardizing CMRI for Post-Surgical Tetralogy of Fallot
Shiraz Maskatia, MD
Tetralogy of Fallot - Background
Figure 6: Factors influencing right ventricular (RV) dysfunction and impaired clinical status after TOF repair.
Tetralogy of Fallot - Background
Right Ventricular Fibrosis

• In adult pts, LV and RV increased LGE associated with:
  - Arrhythmia
  - Exec intolerance
  - RV dysfunction

*Ventricular Fibrosis Suggested by Cardiovascular Magnetic Resonance in Adults With Repaired Tetralogy of Fallot and Its Relationship to Adverse Markers of Clinical Outcome*

Sarka V. Babu-Narayan, BSc, MRCP; Philip J. Kilner, MD, PhD; Wei Li, MD, PhD; James C. Moon, MRCP; Omar Goktekin, MD; Periklis A. Davlouros, MD; Mohammed Khan, MPH; Siew Yen Ho, PhD, FRCP; Dudley J. Pennell, MD, FRCP; Michael A. Gatzoulis, MD, PhD

*Circulation. 2006;113:405-413*
Left Ventricular Dysfunction

Figure 7: Association between right ventricular (RV) and left ventricular (LV) ejection fraction (EF) in 100 patients with repaired TOF [10].

\[ RVEF = 2.26 + 0.76 \times LVEF \]

\[ r = 0.58 \]
\[ p < 0.001 \]

Geva, JACC 2004
## Indications

### Table 1 Structural and functional abnormalities encountered in repaired TOF

<table>
<thead>
<tr>
<th>Structural Abnormalities</th>
<th>Functional Abnormalities</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infundibulotomy scar</td>
<td>Pulmonary regurgitation</td>
</tr>
<tr>
<td>RVOT obstruction</td>
<td>Tricuspid regurgitation</td>
</tr>
<tr>
<td>Branch PA stenosis</td>
<td>RV systolic dysfunction</td>
</tr>
<tr>
<td>RV fibrosis</td>
<td>RV diastolic dysfunction</td>
</tr>
<tr>
<td>Dilated aorta</td>
<td>LV dysfunction</td>
</tr>
<tr>
<td>Associated genetic and non-cardiac anomalies</td>
<td>Neurodevelopmental abnormalities</td>
</tr>
</tbody>
</table>

Geva, JCMR 2011
ACC/AHA Guideline

ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: Executive Summary

A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines for the Management of Adults With Congenital Heart Disease)

Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons

WRITING COMMITTEE MEMBERS
Carole A. Wames, MD, FRCP, FACC, FAHA, Co-Chair; Roberta G. Williams, MD, MACC, FAHA, Co-Chair; Thomas M. Bashore, MD, FACC; John S. Child, MD, FACC, FAHA; Heidi M. Connolly, MD, FACC; Joseph A. Dearani, MD, FACC; Pedro del Nido, MD; James W. Fasules, MD, FACC; Thomas P. Graham, Jr, MD, FACC; Ziyad M. Hijazi, MBBS, MPH, FACC, FSCAI; Sharon A. Hunt, MD, FACC, FAHA; Mary Eta King, MD, FACC, FASE; Michael J. Landzberg, MD, FACC; Pamela D. Miner, RN, MN, NP; Martha J. Radford, MD, FACC; Edward P. Walsh, MD, FACC; Gary D. Webb, MD, FACC

TASK FORCE MEMBERS
Sidney C. Smith, Jr, MD, FACC, FAHA, Chair; Alice K. Jacobs, MD, FACC, FAHA, Vice-Chair; Cynthia Adams, RSN, PhD, FAHA; Jeffrey L. Anderson, MD, FACC, FAHA; Elliott M. Antman, MD, FACC, FAHA; Christopher E. Buller, MD, FACC; Mark A. Creager, MD, FACC, FAHA; Steven M. Ettinger, MD, FACC; Jonathan L. Halperin, MD, FACC, FAHA; Sharon A. Hunt, MD, FACC, FAHA; Harlan M. Krumholz, MD, FACC, FAHA; Frederick G. Kushner, MD, FACC, FAHA; Bruce W. Lytle, MD, FACC, FAHA; Rick A. Nishimura, MD, FACC, FAHA; Richard L. Page, MD, FACC, FAHA; Barbara Riegel, DNP, RN, FAHA; Lynn G. Tarkington, RN; Clyde W. Yancy, MD, FACC, FAHA

*Society of Thoracic Surgeons representative.
**International Society for Adult Congenital Heart Disease representative.
**Society for Cardiovascular Angiography and Interventions representative.
***American Society of Echocardiography representative.
****Heart Rhythm Society representative.
†Canadian Cardiovascular Society representative.
‡Former Task Force member during this writing effort.

This document was approved by the American College of Cardiology Foundation Board of Trustees in July 2008 and by the American Heart Association Science Advisory and Coordinating Committee in August 2008.


This article has been copublished in the Journal of the American College of Cardiology.

Permissions: Multiple copies, modification, alteration, enhancement, and/or distribution of this document are not permitted without the express permission of the American Heart Association. Instructions for obtaining permission are located at http://www.americanheart.org/presenter.jhtml?identifier=303999 by selecting either the “topic list” link or the “chronological list” link (No. LS-1833). To purchase additional reprints, call 833-216-2533 or e-mail ilee@rmay@wolkenskizer.com.

Circulation is available at http://circ.ahajournals.org

DOI: 10.1161/CIRCULATIONAHA.108.190811

© 2008 American College of Cardiology Foundation and American Heart Association, Inc.
Indications-Imaging guidelines

• ACC/AHA practice guidelines for adults with congenital heart disease

• Class I (Level of Evidence: C)
  - Patients with tetralogy of Fallot should have echocardiographic examinations and/or magnetic resonance imagings (MRIs) performed by staff with expertise in ACHD
Indications-Intervention guidelines

• Class 1

- Pulmonary valve replacement is indicated for severe pulmonary regurgitation and symptoms or decreased exercise tolerance. *(Level of Evidence: B)*

- Coronary artery anatomy, specifically the possibility of an anomalous anterior descending coronary artery across the RVOT, should be ascertained before operative intervention. *(Level of Evidence: C)*
Indications-Intervention guidelines

• Class IIa

- Pulmonary valve replacement is reasonable in adults with previous tetralogy of Fallot, **severe pulmonary regurgitation**, and any of the following:

  - Moderate to severe right ventricular (RV) dysfunction. *(Level of Evidence: B)*

  - Moderate to severe RV enlargement. *(Level of Evidence: B)*

  - Moderate to severe tricuspid regurgitation (TR). *(Level of Evidence: C)*
Indications-Intervention guidelines

• Class IIa

- Surgery is reasonable in adults with prior repair of tetralogy of Fallot and residual RVOT obstruction (valvular or subvalvular) and any of the following:

  • Residual RVOT obstruction (valvular or subvalvular) with progressive and/or severe dilatation of the right ventricle with dysfunction. (*Level of Evidence: C*)

  • Residual ventricular septal defect (VSD) with a left-to-right shunt greater than 1.5:1. (*Level of Evidence: B*)

  • Severe aortic regurgitation (AR) with associated symptoms or more than mild LV dysfunction. (*Level of Evidence: C*)

  • A combination of multiple residual lesions (e.g., VSD and RVOT obstruction) leading to RV enlargement or reduced RV function. (*Level of Evidence: C*)
Why do I do it?

• Right Ventricle
  - Size and Function
  - Recurrent/Residual RVOT obstruction
  - Regional wall motion abnormalities
    • Assessment of myocardial viability?
  - Pulmonary regurgitation
  - Tricuspid Regurgitation
Why do I do it?

• Pulmonary Arteries
  - Branch PA stenosis
  - Differential Flow/Regurgitation
Why do I do it?

• Left ventricle
  - Size and systolic function

• Aortic Root
  - Size
  - Insufficiency

• Aortic arch
  - Sidedness, branching pattern
Why do I do it?

• Presence/Absence of AP collaterals

• Coronary artery branching pattern
Our Protocol

• BB: axial (T1EPI)

• Cine
  - SSFP 4C-cover heart, SA, VLA (ensure RV and LV), LVOT (3 slices), RVOT (3 slices)
  - Cine imaging PAs
  - SSFP aortic root short axis (3-5 slices)

• Phase contrast:
  - Aorta (STJ)
  - MPA
  - Branch Pas
  - AVVs

• MRA

• 3D SSFP
  - axial for coronaries
  - only on initial studies

• no LGE