

**Best Evidence Statement (BESt)**

August 28, 2010

**Follow-up Testing After Tetralogy of Fallot Repair**
**Clinical Question**

- P (population/problem) Among patients age 1 year through adult with repaired tetralogy of Fallot (TOF)
- I (intervention) what is the optimum frequency of:
- office visit
  - electrocardiogram (ECG)
  - chest x-ray
  - scale of function / symptomatology
  - echocardiogram
  - magnetic resonance imaging (MRI)
  - exercise testing
  - Holter monitor
- O (outcome) to help detect risk of sudden death, residual pulmonary stenosis, residual pulmonary regurgitation, right ventricular failure, arrhythmia, or quality of life concerns?

**Target Population**

Patients with repaired TOF ages 1 year through adult

**Recommendation** (See Table of Recommendation Strength following references)

It is recommended that patients, age 1 year through adult with repaired TOF be evaluated periodically according to the following schedule:

	<b>Begin</b>	<b>Repeat</b>
<b>Office visit</b>	Age 1	Annually
<b>ECG</b>	Age 1	Annually
<b>Chest x-ray</b>	Age 1	Every 5 years
<b>NYHA functional scale classification</b> <a href="#">See appendix</a>	Age 3	Annually
<b>Echocardiogram / MRI</b> (frequency after age 10 is dependent on type)		
a. patients with repaired tetralogy of Fallot with more than moderate pulmonary regurgitation (PR) <sup>1, 2</sup>		
echocardiogram <sup>2</sup>	Age 1	Annually until age 10 then every 2 years
MRI <sup>2</sup>	Age 9 or 10	Every 2 years
b. all other TOF patients <sup>3</sup>		
echocardiogram <sup>3</sup>	Age 1	Annually until age 9 or 10 then 2 out of 3 years
MRI <sup>3</sup>	Age 9 or 10	Every 3 years
<b>Exercise testing</b>	Age 9 or 10	Every 3 years
<b>Holter monitor</b>	Age 6 or 7	Every 3 years

Abbreviations: ECG = electrocardiogram; MRI = magnetic resonance imaging; NYHA = New York Heart Association; TOF = tetralogy of Fallot  
(*Local Consensus [5], Geva 2006 [5b]*)

<sup>1</sup> “more than moderate PR” defined as either moderate or greater degree of PR by echo, or PR regurgitant fraction > 20% by MRI  
<sup>2</sup> alternate echocardiogram and MRI each year for routine monitoring (age 10 and older) for patients with repaired tetralogy of Fallot with moderate or greater degree of PR, the most common type of TOF defect  
<sup>3</sup> all other patients may follow a schedule of MRI once every 3 years with an echocardiogram on the other two years (age 10 and older)

## Discussion/summary of evidence

The body of evidence is low quality. There is one review article that discusses assessment testing and frequency (*Geva 2006 [5b]*). Local consensus is in agreement with much of the information in the article as outlined in the table above (*Local Consensus [5], Geva 2006 [5b]*).

## Health Benefits, Side Effects and Risks

The goals of diagnostic evaluation in patients with repaired TOF include identification of structural and functional abnormalities, evaluation of the severity of the hemo-dynamic abnormalities, surveillance of the rate and degree of disease progression and identification of markers of poor prognosis (*Geva 2006 [5b]*). The data derived from the assessments and laboratory tests guide clinical decisions (*Geva 2006 [5b]*). The benefit to following these recommendations is early identification of pulmonary regurgitation in order to achieve the outcomes listed in the clinical question and to guide decisions for timing of pulmonary valve replacement.

There is minimal risk for each of the non-invasive evaluation tests and procedures; an annual chest x-ray includes a low radiation exposure. Though an MRI is minimally invasive, risk increases if contrast or sedation are used, or if it is inappropriately administered to patients with implanted metal devices. MRI's may confer burden to the patient and the system due to its cost.

## References (evidence grade in [ ]; see *Table of Evidence Levels following references*)

**Note:** When using the electronic version of this document,  indicates a hyperlink to the PubMed abstract. A hyperlink following this symbol goes to the article PDF when the user is within the CCHMC network.

1. **Geva, T.:** Indications and timing of pulmonary valve replacement after tetralogy of Fallot repair. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu:* 11-22, 2006, [5b]  .
2. **Local Consensus:** During recommendation development timeframe. [5] .

Note: Full tables of evidence grading system available in separate document:

- [Table of Evidence Levels of Individual Studies by Domain, Study Design, & Quality](#) (abbreviated table below)
- [Grading a Body of Evidence to Answer a Clinical Question](#)
- [Judging the Strength of a Recommendation](#) (abbreviated table below)

**Table of Evidence Levels** (see note above)

<i>Quality level</i>	<i>Definition</i>
1a† or 1b†	Systematic review, meta-analysis, or meta-synthesis of multiple studies
2a or 2b	Best study design for domain
3a or 3b	Fair study design for domain
4a or 4b	Weak study design for domain
5a or 5b	Other: General review, expert opinion, case report, consensus report, or guideline

†a = good quality study; b = lesser quality study

**Table of Recommendation Strength** (see note above)

<b>Strength</b>	<b>Definition</b>
“Strongly recommended”	There is consensus that benefits clearly outweigh risks and burdens (or visa-versa for negative recommendations).
“Recommended”	There is consensus that benefits are closely balanced with risks and burdens.
No recommendation made	There is lack of consensus to direct development of a recommendation.
<b>Dimensions:</b> In determining the strength of a recommendation, the development group makes a considered judgment in a consensus process that incorporates critically appraised evidence, clinical experience, and other dimensions as listed below.	
<ol style="list-style-type: none"> <li>1. Grade of the Body of Evidence (see note above)</li> <li>2. Safety / Harm</li> <li>3. Health benefit to patient (<i>direct benefit</i>)</li> <li>4. Burden to patient of adherence to recommendation (<i>cost, hassle, discomfort, pain, motivation, ability to adhere, time</i>)</li> <li>5. Cost-effectiveness to healthcare system (<i>balance of cost / savings of resources, staff time, and supplies based on published studies or onsite analysis</i>)</li> <li>6. Directness (<i>the extent to which the body of evidence directly answers the clinical question [population/problem, intervention, comparison, outcome]</i>)</li> <li>7. Impact on morbidity/mortality or quality of life</li> </ol>	

## Supporting information

### Introductory/background information

The surgical repair of TOF leaves a child or adolescent with many major anatomical and functional abnormalities that are often well tolerated. Pulmonary regurgitation is one such anomaly that develops and worsens over the years post TOF repair, subsequently requiring pulmonary valve replacement (*Geva 2006 [5b]*). It is the data from the assessments and diagnostic evaluations that guide clinical decisions including the indication for and timing of pulmonary valve replacement (*Geva 2006 [5b]*).

### Group/team members

Tom Kimball, MD, Heart Institute  
 Bill Gottliebson, MD, Heart Institute  
 Erik Michelfelder, MD, Heart Institute  
 Gary Webb, MD, Heart Institute  
 Jean Giver, RN, Heart Institute  
 Catherine Krawczeski, MD, Heart Institute

#### *Clinical Effectiveness Support Personnel:*

Carla Williams, MSA, Outcomes Manager  
 Anjali Basu, MS, Associate Outcomes Manager  
 Eloise Clark, MPH, MBA, Guidelines Program Administrator, Evidence Facilitator  
 Danette Stanko-Lopp, MA, MPH, Epidemiologist  
 Karen Vonderhaar, MS, RN, Guidelines Program Administrator, Methodologist

### Search strategy

#### Original Search

OVID Databases: MedLine, CINAHL, Cochrane, and Other – Medical Association and Guideline Websites: American Academy of Pediatrics (AAP), American College of Cardiology (ACCF), American Academy of Neurology (AAN), American College of Emergency Physicians, American Heart Association (AHA), European Society of Cardiology, National Guideline Clearinghouse (HGC)  
 Search Term & MeSH Term – exp Tetralogy of Fallot

#### Limits:

- English language
- 1996 to April, 2010
- All ages

- (guideline or meta analysis or practice guidelines or systematic review).pt. or "the cochrane library".jn. or "cochrane database of systematic reviews".jn.

**Applicability issues** – Proposed process and outcome measures include:

Percentage of patients with repaired tetralogy of Fallot who had an annual clinic visit.

Percentage of patients with repaired tetralogy of Fallot who had an annual electrocardiogram conducted.

Percent of patients with repaired tetralogy of Fallot who had a chest x-ray conducted every 5 years.

Percent of patients with repaired tetralogy of Fallot, greater than or equal to 10 years of age, with moderate or greater degree of pulmonary regurgitation (PR), defined as moderate or greater by echo or PR regurgitant fraction > 20% by MRI.

Copies of this Best Evidence Statement (BESt) are available online and may be distributed by any organization for the global purpose of improving child health outcomes. Website address: <http://www.cincinnatichildrens.org/svc/alpha/h/health-policy/ev-based/default.htm>

Examples of approved uses of the BESt include the following:

- copies may be provided to anyone involved in the organization's process for developing and implementing evidence based care;
- hyperlinks to the CCHMC website may be placed on the organization's website;
- the BESt may be adopted or adapted for use within the organization, provided that CCHMC receives appropriate attribution on all written or electronic documents; and
- copies may be provided to patients and the clinicians who manage their care.

Notification of CCHMC at [HPCEInfo@cchmc.org](mailto:HPCEInfo@cchmc.org) for any BESt adopted, adapted, implemented or hyperlinked by the organization is appreciated.

For more information about CCHMC Best Evidence Statements and the development process, contact Health Policy & Clinical Effectiveness office at: 513-636-2501 or [HPCEInfo@cchmc.org](mailto:HPCEInfo@cchmc.org).

**Note**

**This Best Evidence Statement addresses only key points of care for the target population; it is not intended to be a comprehensive practice guideline. These recommendations result from review of literature and practices current at the time of their formulation. This Best Evidence Statement does not preclude using care modalities proven efficacious in studies published subsequent to the current revision of this document. This document is not intended to impose standards of care preventing selective variances from the recommendations to meet the specific and unique requirements of individual patients. Adherence to this Statement is voluntary. The clinician in light of the individual circumstances presented by the patient must make the ultimate judgment regarding the priority of any specific procedure.**

**Reviewed against quality criteria by 2 independent reviewers.**

## Appendix

The **New York Heart Association (NYHA) Functional Classification** provides a simple way of classifying the extent of [heart failure](#). It places patients in one of four categories based on how much they are limited during physical activity; the limitations/symptoms are in regards to normal breathing and varying degrees in shortness of breath and or angina pain:

NYHA Class	Symptoms
I	No symptoms and no limitation in ordinary physical activity, e.g. shortness of breath when walking, climbing stairs etc.
II	Mild symptoms (mild shortness of breath and/or angina) and slight limitation during ordinary activity.
III	Marked limitation in activity due to symptoms, even during less-than-ordinary activity, e.g. walking short distances (20–100 m). Comfortable only at rest.
IV	Severe limitations. Experiences symptoms even while <b>at rest</b> . Mostly bedbound patients