

# CONGENITAL CARDIOLOGY TODAY

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**PICS-CSI Asia 2016 - Catheter Interventions in Congenital, Structural and Valvular Heart Disease**  
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**The 13<sup>th</sup> International Dead Sea Symposium (IDSS) on Innovations in Cardiac Arrhythmias and Device Therapy**  
Mar. 6-9, 2016; Tel Aviv, Israel  
<http://idss-ep.com>

**Congenital and Paediatric Echocardiography From Basics to Advanced**  
Apr. 28-30, 2016; Toronto, On Canada  
[www.cvent.com/d/zfjx4](http://www.cvent.com/d/zfjx4)

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## **Surgical Outcomes in Pediatric Patients, Who Underwent Tetralogy of Fallot Correction, Relative to Different Techniques Used to Relieve Right Ventricular Outflow Obstruction**

By Lorieilyn G. Mandigma, MD (Investigator); Ma. Bernadette A. Azcueta, MD (Co-investigator); Corazon A. Estevanez, MD (Consultant Adviser); Mercilyn Yap, MD (Consultant Adviser)

### **Introduction**

Tetralogy of Fallot (TOF) is the most common Cyanotic Congenital Heart Disease, with an incidence of approximately 0.5/1000 live births (5% to 7% of congenital heart lesions).<sup>1</sup> Before the advent of surgical intervention, about 50% of patients with Tetralogy of Fallot died in the first few years of life, and it was unusual for a patient to survive more than 30 years.<sup>2</sup> Most patients died in childhood with a rate of survival of 66% at 1 year of age, 40% at 3 years, 11% at 20 years and 3% at 40 years.<sup>1,2</sup> Nowadays, with the advent of surgical repair, which includes closure of the Ventricular Septal Defect (VSD) and relief of Right Ventricular Outflow Tract (RVOT), the long-term survival of TOF patients has been greatly improved.<sup>1</sup>

Nevertheless, a myriad of potential complications have been reported in operated TOF patients that underlie the importance of follow-up after surgery, even if they remain asymptomatic.<sup>1</sup> These complications include rhythm and conduction disorders such as: sudden cardiac deaths, pulmonary regurgitation (PR) with RV dilatation and dysfunction,

residual RVOT obstruction. These complications may lead to subsequent hospitalizations, repeat operations, arrhythmias, and heart failure, as well as death following initial corrective surgery.<sup>1</sup>

These relevant postoperative residues bring about limitation in the right ventricular function and, accordingly, the quality of life and life expectancy.

### **Review of Related Literature**

Reparative surgery for TOF should ideally result in complete closure of the VSD, preservation of right ventricular form and function, with an unobstructed right ventricular outflow tract incorporating a competent pulmonary valve.<sup>3</sup> Successful repair through a right ventriculotomy was first achieved by Lillehei and Varco, using "controlled cross-circulation" in 1954.<sup>4</sup> Kirklin et al. was the first to use a pump oxygenator for the repair of TOF 1 year later.<sup>5</sup> Improvements in cardiopulmonary bypass technology, as well as in surgical technique and perioperative care made early repair feasible with low morbidity and mortality.<sup>6</sup> Transatrial/transpulmonary repair of TOF, which was first reported in 1963 by Hudspeth et al., has been an important step in the evolution of TOF surgery.<sup>7</sup> It was reintroduced by Edmunds et al. in 1976, and popularized in recent years. The benefits of the transatrial/transpulmonary approach are believed to derive from eliminating a right

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ventriculotomy, which may lead to late right ventricular (RV) dilatation and dysfunction, as well as increased risk of ventricular ectopic activity.<sup>8</sup> At our center, Tetralogy of Fallot correction has been performed since 1975 when the center became operational. Since 1980, a local study by Bote-Nunez et al., has shown that both transannular patch and valve-sparing techniques are already being performed, and have had excellent late survival with only three known late deaths due to cardiac-related causes.<sup>9</sup> Subjects included in the study belong to functional Class I and II. Residual pulmonary stenosis was also noted on 2D echocardiogram in 60% of the subjects.<sup>9</sup>

A study by Murphy et al. on the long-term outcome in patients undergoing surgical repair of Tetralogy of Fallot showed an overall survival rate of 95% 5 years after operation, 92% after 10 years and 15 years, 91% after 20 years, 87% after 25 years and 86% after 32 years.<sup>10</sup> According to age at the time of operation: 30-year survival was 90% for less than 5 years old, 93% for 5 to 7 years old, 91% for 8 to 11 years old, and 76% for more than 12-years-old.<sup>10</sup> According to patching of pulmonary annulus, 30-year survival was 87% for those without pulmonary outflow patch, 85% for those who had patching through the annulus, and 88% for those who had patching up to the annulus.<sup>10</sup>

Despite the high number of favorable outcome as shown in several studies, several potential complications have been identified and warrant the need for long-term follow-up.

In the study by Cardoso et al., restrictive RV physiology was seen on follow-up of most patients.<sup>11</sup> Incidence varied from 50%-70% in the study by Gatzolius et al. Restrictive physiology increases the chance of ventricular arrhythmia and sudden cardiac death.<sup>12</sup>

Pulmonary regurgitation (PR) complicating surgical repair of TOF is common in all patients, with over 80% having at least moderate-to-severe PR. PR has been shown to be related to the use of a transannular patch during RVOT reconstruction and aggressive infundibulectomy involving the pulmonary valve annulus. The adverse effects of PR include: progressive dilatation of RV, reduced exercise capacity, arrhythmia and sudden death.<sup>13</sup>

Impaired exercise capacity after complete repair of Tetralogy of Fallot is directly related to the degree of residual pulmonary regurgitation.<sup>14</sup>

## Significance of the Study

The aim of this study was to provide current data on the surgical outcome of Tetralogy of Fallot patients who underwent repair at our institution. It also aimed to help establish a guideline on post-operative monitoring and follow-up.

## General Objective

To determine the outcome of pediatric patients who underwent Tetralogy of Fallot correction in relation to the surgical technique used in relieving RVOT.

## Specific Objectives

1. To determine the surgical techniques used in relieving right ventricular outflow tract obstruction (RVOT).
2. To determine the outcome of patients who underwent Tetralogy of Fallot correction in terms of:
  - Presence and degree of pulmonary regurgitation
  - Presence and degree of residual pulmonary stenosis
  - Presence of right ventricular dilatation
  - Presence of RV dysfunction
    - Systolic
    - Diastolic
  - Functional capacity

3. To determine the relationship of the surgical technique used in RVOT repair relative to the determined outcome.

## Hypothesis

### *Null Hypothesis:*

The surgical technique used in relieving RVOT obstruction has no association in the development of unfavorable outcomes in patients who underwent Tetralogy of Fallot correction.

### *Alternative Hypothesis:*

The surgical technique used in relieving RVOT obstruction has an association in the development of unfavorable outcomes in patients who underwent Tetralogy of Fallot correction.

## Methodology

### **Study Design**

Prospective Cohort

### **Study Setting and Period**

Recruitment of subjects for the study took place at the Philippine Heart Center from November 2012 to October 2014.

### **Inclusion and Exclusion Criteria**

#### Inclusion Criteria

- Tetralogy of Fallot patients whose age at the time of surgery was 3-18 years old

#### Exclusion criteria

- TOF with pulmonary valve atresia
- TOF patients with associated Complex Congenital Heart Disease
- TOF patients with genetic abnormalities
- Tetralogy of Fallot patients who had clinical signs of right-sided failure (jaundice, hepatomegaly, pulmonary venous congestion) prior to surgery.
- Tetralogy of Fallot patients who had echocardiographic findings of RV dysfunction prior to surgery.

### **Sample Size**

The sample size computed is  $n = 45$  based on 95% confidence level, maximum tolerable error of 10%, assumed rate of RV diastolic dysfunction of 72.5%,<sup>14</sup> and assumed census of TOF correction.<sup>11</sup>

### **Identification of Variables**

#### **Definition of Terms**

1. **Tetralogy of Fallot** – comprises a constellation of cardiac findings that share the following common anatomic abnormalities: large malaligned VSD, overriding of the aorta over the septal defect, right ventricular outflow obstruction, and right ventricular hypertrophy.
2. **Dyspnea** – difficulty breathing, observed as an increase in respiratory rate and the use of accessory muscles in breathing.
3. **RV Diastolic Dysfunction** – shall be defined echocardiographically as the presence of any of the following signs of diastolic dysfunction: a.) presence of prolonged isovolumic relaxation time, b.) E/A reversal, d) E/A ratio  $>1.5$ , e.)  $E/Ea > 10$ , or f.) presence RV restrictive physiology which

is the presence of laminar antegrade diastolic PA flow throughout the respiratory cycle, which is coincident with atrial systole on 2D echocardiography.

4. **RV Systolic Dysfunction** – shall be defined. echocardiographically as an impairment in any of the following parameters: TAPSE, TDI systolic velocity, or RVEF.
5. **Functional Capacity** – defined as the individual's capacity to perform submaximal activities as measured by the distance walked in six minute exercise test and therefore the ability for patients to perform day-to-day activities.

#### Study Maneuver

After approval of the IERB, eligible subjects for the study were recruited, informed consent was secured from parents/guardians of all subjects; assent form was also secured for patients 7-18 years old. The eligible subjects were further grouped depending on the surgical technique used in relieving their RVOT obstruction (Group A – TOF patients who underwent transannular patching, and Group B – TOF patients who underwent valve-sparing procedure). The demographic characteristics, pre-operative clinical data (hematocrit, oxygen saturation, pulmonary arteries and pulmonary valve Z-score, previous palliative procedure) were obtained. The presence and degree of residual pulmonary stenosis, presence and degree of pulmonary regurgitation, right ventricular dilatation, RV dysfunction (systolic and diastolic) were determined by echocardiographic assessment prior to hospital discharge of subject (at least one week, up to one month post-operation). Transthoracic 2-dimensional echocardiography was performed and interpreted on the eligible subjects by three echocardiographers to avoid bias. RV diastolic dysfunction was determined by the presence of any of the following signs of diastolic dysfunction by 2D echocardiography:

- a) Presence of shortened isovolumic relaxation time,
- b) E/A reversal,
- c) E/A ratio >1.5,
- d) E/Ea > 10, or
- e) Presence RV restrictive physiology.

Determination was done by M-mode imaging of the right ventricular cavity, followed by detailed pulsed Doppler echocardiography: tricuspid and characteristics at the level of the tips of the valve leaflets in apical 4-chamber view. The antegrade diastolic PA flow, E:A ratio, and right ventricular isovolumic relaxation time was measured. Tissue Doppler imaging was used to measure RV diastolic dysfunction. For RV systolic function determination, the parameters that were determined were RV ejection fraction, and tricuspid annular plane systolic excursion (TAPSE). Tricuspid annular plane excursion was used to arrive at a quantitative measurement of RV systolic function by directing the M-Mode cursor from the apex to the medial tricuspid annulus during systole or when RV shortens from base to apex. Tissue Doppler imaging was also used to measure RV systolic dysfunction.

The Six-Minute Walk Test (6MWT) was used to predict the functional capacity of the subjects. Functional status classification was done using the Modified Ross classification for heart failure in children. The Six-Minute Walk Test was done on OPD follow-up (at least two weeks post-operation). For subjects aged three to six years old, psychological conditioning regarding Six-Minute Walk Test was done prior to hospital



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Table 1 – Pre-operative Clinical Data of Patients Who Underwent TOF Correction			
	*Group A N = 23 (%)	**Group B N = 40 (%)	P value
<b>Sex</b>			1
Male	13 (56.5%)	23 (57.5%)	
Female	10 (43.5%)	17 (42.5%)	
<b>Age (years + SD)</b>	7.5 + 3.5	7.0 + 3.8	0.64
<b>Weight (kg + SD)</b>	22.5 + 10.64	19.5 + 9.89	0.27
<b>Hematocrit (SI unit + SD)</b>	0.56 + 0.08	0.53 + .09	0.27
<b>Oxygen saturation (%+ SD)</b>	77.7 + 7.0	77.6 + 7.4	0.94
<b>Previous palliative surgery</b>			0.13
Yes	3 (13%)	1 (2%)	
No	20 (87%)	39 (98%)	
<b>Z score</b>			
Right pulmonary artery (z score + SD)	-0.95 + 0.93	0.79 + 0.86	0.49
Left pulmonary artery (z score + SD)	-1.1 + 1	- 0.75 +0.78	0.17
Pulmonary valve annulus (z score + SD)	- 2.5 + 1.7	- 2.1 + 1.1	0.27
* Group A – Transannular Patch Group			
**Group B – Valve-sparing Group			

discharge, and prior to the test on follow-up. If any of the subjects, regardless of their age and reason, failed to finish the test, he/she was rescheduled at another time within a month, until he/she was able to complete the test. The data that were determined were: distance walked in six minutes, oxygen saturation and heart rate during the six minutes and during a three-minutes recovery period. The patient's functional status was assessed using the Modified Ross classification for heart failure in children, which was done through patient and parent/guardian interview.

#### Plan for Analysis

Continuous variables were calculated as mean + standard deviation or median and range. Significance was determined at a p value of < 0.05. All p values are two-sided, and confidence intervals are 95%. The two groups were compared by chi-square tests for dichotomous variables and student's t tests for continuous variables.

#### Ethical Considerations

The study was conducted in compliance with the ethical principles set forth in the Declaration of Helsinki. Prior to the study initiation, a review and approval of the study protocol and informed consent and subsequent amendments by the Philippine Heart Center Institutional Ethics Review Board (PHC IERB) was done.

Before a subject's participation, a written informed consent was obtained by the investigator after adequate explanation of the aims, methods, anticipated benefits, and potential risks of the study. The informed consent was signed and personally dated by the subject, and the person who conducted the informed consent discussion. One copy of the informed consent was given to the subject.

The investigator preserved the confidentiality of all subjects taking part in the study. The investigator ensured that the subject's anonymity was maintained.

#### Results

Table 1 shows the baseline characteristics of the two groups being compared in this study. Results show that the subjects in Group A were older and had a greater weight compared to Group B, although

Table 2 – Post-Operative Outcome of Patients Who Underwent TOF Correction			
	*Group A N = 23 (%)	**Group B N = 40 (%)	P value
<b>Presence and degree of pulmonary regurgitation</b>			0.798
None	16 (69.6%)	26 (65%)	
Mild	7 (30.4%)	11 (27.5%)	
Moderate	0 (0%)	0 (0%)	
Severe	0 (0%)	0 (0%)	
<b>Presence and degree of residual pulmonary stenosis</b>			0.229
Mild	23 (100%)	20 (50%)	
Moderate	0 (0%)	15 (37.5%)	
Severe	0 (0%)	5 (12.5%)	
<b>Presence of right ventricular dilatation</b>			1.0
Yes	14 (60.9%)	24 (60%)	
None	9 (39.1%)	16 (40%)	
<b>Presence of RV dysfunction</b>			0.6
Systolic			
Yes	13 (56.5%)	19 (47.5%)	
None	10 (43.5%)	21 (52.5%)	
Diastolic			0.51
Yes	21 (91.3%)	34 (85%)	
None	2 (8.7%)	6 (15%)	
<b>Distance travelled in six minutes (meters + SD)</b>	297 + 71.3	215.3 + 69.2	0.81
* Group A – Transannular Patch Group			
**Group B – Valve-sparing Group			

Table 3 – Functional Assessment by Modified Ross Classification for Heart Failure in Children in Patients Who Underwent TOF Correction			
	*Group A N = 23 (%)	**Group B N = 40 (%)	P value
<b>Class I</b>	0	0	1.0
<b>Class II</b>	22 (96.7%)	39 (97.5%)	
<b>Class III</b>	1 (4.3%)	1 (2.5%)	
<b>Class IV</b>	0	0	
* Group A – Transannular Patch Group			
**Group B – Valve-sparing Group			

there was no significant difference between the two groups.

Table 2 shows the outcome of patients in the two groups during the immediate post-operative period. Results showed that post-operatively, pulmonary regurgitation was more common among the transannular patch group (Group A) at 30.4% compared to the valve-sparing group (Group B) at 27.5%. All patients who had pulmonary regurgitation in both groups have mild severity.

In terms of residual pulmonary stenosis, all patients in both groups had residual pulmonary stenosis. Patients from the valve-sparing group (Group B) had greater severity, with 15 having moderate pulmonary stenosis, and 5 having severe pulmonary stenosis. This finding, however, did not yield a statistical difference.

Right ventricular dilatation was still present on most patients on both groups, with Group A at 60.9% and Group B at 60%.

RV systolic dysfunction was more common in Group A patients, accounting for 56.5% of the group, while in Group B, it was present in 19 patients, accounting for 25% of the group.



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RV diastolic dysfunction was present in 91.3% of Group A and 85% in Group B. Specifically, Stage 3 diastolic dysfunction, which connotes restrictive physiology, is present in 33% of the patients in Group A and 20% in Group B.

With regards to the distance travelled in 6 Minute Walk Test, Group A has a longer distance travelled at a mean of 297 + 71.3m as compared to Group B, at 215.3 + 69.2m.

**Table 3** shows the functional classification of the patients who underwent Tetralogy of Fallot correction. Most patients in Group A (96.7%) are in functional Class II, while there was only 1 (4.3%) who was in functional Class III. This patient had a prolonged hospital stay due to chylothorax and right ventricle failure. This patient was subsequently diagnosed with Absent Left-Pulmonary Artery on hemodynamic studies that were done postoperatively. In Group B, 97.5% were in functional Class II while one patient was in functional Class III. This patient had previous Blalock-Taussig Shunt (BTS) surgery, which was patent, but was not able to be taken down intraoperatively due to technical difficulties. This patient subsequently had a reoperation after 8 months, and was subsequently discharged; however, the patient apparently died after 4 months.

All the results that were gathered did not yield any statistical significance.

## Discussion

Surgical repair by means of Ventricular Septal Defect (VSD) closure and relief of right ventricular outflow obstruction is the definitive management for Tetralogy of Fallot. Over the years, there has been consistent progress in refining the surgical techniques in improving anesthetic and critical care management to achieve this goal. This evolution has provided better postoperative outcomes and has led to improvement in the quality of life of repaired TOF patients as they reach adulthood.

Postoperative complications of pulmonary incompetence and right ventricular dysfunction can be attributed to many factors, among which, the surgical technique used to relieve right ventricular outflow tract dysfunction has been implied as one of the many causes.

In our study, pulmonary incompetence was already seen among patients in both groups, with the transannular group having a greater incidence at 30.4%, as compared to the pulmonary valve-sparing group at 27.5%. Both groups had mild severity. Not long ago, pulmonary incompetence was regarded as an inevitable, but unimportant late sequelae of repair. Much emphasis was placed on the need for complete relief of obstruction, often at the expense of a freely regurgitant and ever-dilating outflow tract.<sup>3</sup> These misguided assertions of investigators can be understood when the time course of the effects of postoperative pulmonary incompetence is considered. Problems from pulmonary incompetence however occur decades after repair,<sup>3</sup> and is due to progressive RV dilatation and pressure overload.

Both groups had residual pulmonary stenosis. All patients in the transannular group had mild severity. In the pulmonary valve sparing group, 50% had mild, 7.5% had moderate, and 2.5% had severe severity. In the past 20 years, a shift from the need for complete relief of obstruction<sup>15</sup> towards a policy to preserve the pulmonary valve, even at the expense of a modest residual stenosis, has occurred.<sup>16</sup> This shift might keep adverse late effects of pulmonary incompetence to a minimum, and retain the integrity of the outflow tract, avoiding late aneurysmal dilation.<sup>3</sup>

Findings of both diastolic and systolic RV dysfunction were noted at the early post-operative period, of which, there were more patients in the transannular patching group who had diastolic dysfunction (91.3%), compared to the valve-sparing group (85%). In the study of Cardoso and Miyague,<sup>11</sup> RV restrictive physiology, which connotes diastolic dysfunction, was seen in 63.3% of their patients who underwent Tetralogy of Fallot correction. In their particular study, most of their subjects underwent transannular patching. Authors of previous

studies<sup>17,18,19</sup> have found a correlation between this technique (transannular patching) and the appearance of antegrade diastolic flow in the pulmonary artery, which is seen in 2D echocardiography of patients with restrictive physiology. Restrictive right ventricular physiology can occur both early and late after repair. Norgard et al. found in their study that the anatomical substrate requiring a transannular patch repair is the most important determinant for early restriction, and mid-term restriction is most likely to occur if early restriction is present.<sup>17</sup> Restrictive physiology may relate to a process of endomyocardial fibrosis intrinsic to the disease itself, and intensify as time goes by;<sup>20</sup> to the ventriculotomy; and to the interposition of patches in the interventricular septum and in the infundibulum.<sup>17</sup> It also seems to be influenced by immaturity of the right ventricle, which adapts to the aggressions of extracorporeal circulation, cardioplegia, and hypothermia.<sup>21</sup> In the study done by Cardoso and Miyague,<sup>11</sup> they have concluded that early postoperative restrictive right ventricular physiology (diastolic dysfunction) may be a transient phenomenon of incomplete adaptation of that ventricle to volume and pressure modifications.

RV systolic dysfunction was more common in the pulmonary valve-sparing group at 52.5% compared to the transannular group at 43.5%. These findings were contrary to the findings of Nair et al, wherein, RV dysfunction was more common in patients who needed a transannular patch. They also found that this remarkable decrease in systolic wall motion velocity occurred despite the presence of volume overload in all of their patients, because of the concomitant pulmonary regurgitation.<sup>22</sup> This difference may be explained by the timing of follow-up. Findings in our study were during the early post-operative period as compared to the study of Nair et al, wherein, their findings were noted at least 10 years post-op.

Almost all patients in both groups belong to functional Class II. This finding is expected to improve after full recovery from surgery. There was only one patient from both groups who was in functional Class III. This patient from the transannular group had systolic and diastolic RV dysfunction postoperatively, and suffered from RV failure. He needed prolonged inotropic support and several failure medications to relieve his symptoms. Subsequent cardiac catheterization data revealed that an absent left pulmonary artery was the etiology of the patient's RV failure. In the pulmonary valve sparing group, the patient who was in functional Class III was already 12-years-old, and had undergone previous palliative surgery. Technical difficulties were encountered during her surgery, and the patent shunt from the previous palliative surgery was not removed. Postoperatively, she had RV systolic dysfunction, and was noted to have a significant VSD leak causing volume overload and pulmonary congestion.

Results of the Six-Minute Walk Test for both groups were sub-optimal at a mean of 297 + 71.3m for Group A, and 215.3 + 69.2m for Group B as compared to the normal value by age which is 470 + 57m.<sup>23</sup> Like functional class, these results are expected to improve after full recovery from surgery.

The results of this study did not show statistical difference between the two groups as compared to previously published studies, but it has confirmed the presence of RV dysfunction in the early postoperative period. The implications of these findings suggest a well-catalogued and continuous follow-up of repaired TOF patients. As shown in the study by Remotigue et al., both RV systolic and diastolic dysfunction were present among repaired TOF patients after one to two years of follow-up, and transannular patching technique was associated with the occurrence of restrictive physiology. These findings show that complications at the early postoperative period, although they may be a transient phenomenon, may still persist during the midterm follow-up.

## Limitations of the Study

Most subjects of this study were preoperatively risk-stratified as low risk. There were only 13 patients who had additional risk factors for surgery.

Ten of which were adolescents, and 4 of which underwent previous palliative surgery.

This variable may have affected the findings of this study, since the results may not be a true reflection of the TOF patients undergoing TOF correction.

### Conclusion

Both RV systolic and diastolic dysfunction were present in both groups in the early postoperative period. Diastolic dysfunction was more common among patients who had transannular patching, while systolic dysfunction was more common among patients who had pulmonary valve sparing.

Our study showed that the surgical technique used to relieve the right ventricular outflow obstruction, is not, by itself, associated with the development of right ventricular dysfunction.

Pulmonary incompetence was a more common finding among patients who underwent transannular patching.

Most patients in both groups were in functional Class II, and had sub-optimal distance travelled in Six-Minute Walk Test.

### Recommendations

1. Vigilant monitoring of those patients diagnosed to have right ventricular dysfunction in order to address its impact on the patient's recovery.
2. Post-op Tetralogy of Fallot patients must continuously be followed-up to check for the persistence of RV dysfunction and the evolution of pulmonary incompetence.
3. A prospective study that would equally include TOF patients with pre-operative moderate and high-risk stratification per group, should be done in order to evaluate the true incidence of right ventricular dysfunction among post-op Tetralogy of Fallot patients.

### Appendix A

#### Modified Ross Heart Failure Classification for Children

**Class I** Asymptomatic

**Class II** Mild tachypnea or diaphoresis with feeding in infants.  
Dyspnea on exertion in older children.

**Class III** Marked tachypnea or diaphoresis with feeding in infants.  
Marked dyspnea on exertion.  
Prolonged feeding times with growth failure.

**Class IV** Symptoms such as: tachypnea, retractions, grunting, or diaphoresis at rest.

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## OPPORTUNITY IN PEDIATRIC CARDIOLOGY - MRI CORPUS CHRISTI, TEXAS

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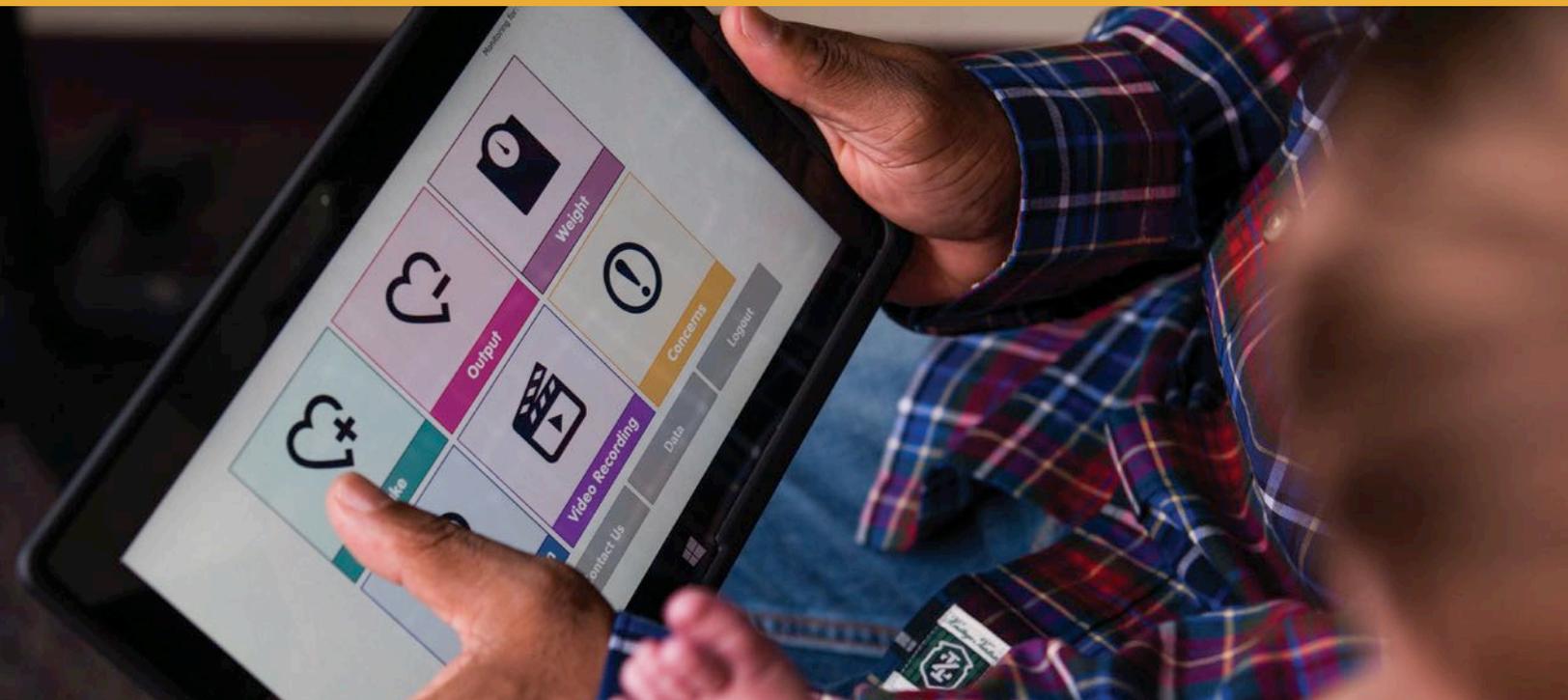
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# Tools to Stay Organized: Using Software to Enhance Efficiency

By Animesh (Aashoo) Tandon, MD, MS

**Keywords:** Software, Mobile, Technology

## Introduction

The digital age has brought significant changes to physicians' lives, both personally and professionally. The ways we interact with patients and make clinical decisions have been strongly influenced by the vastly increased amount and availability of information. Many physicians carry a powerful computer in their hands in the form of a smartphone, which allows nearly instant access to medical journals, textbooks, and guidelines.<sup>1, 2</sup> The ubiquity of social media has also required adaptations of the way that physicians and patients interact.<sup>3</sup> Smartphones are also showing utility in replacing the pager as the preferred method of communication in healthcare settings.<sup>4, 5</sup>

In addition to influencing patient care and communication, technology can also be beneficial in staying organized and efficient; software tools that can be used across devices and platforms are one such example. A great time to experiment with these tools is during fellowship training, so that each individual can find the combination of tools best suited for her or his needs early on. The goal of this article is to outline three categories of software programs that, individually and especially in combination, can yield significant time savings for early career physicians. These programs can especially be beneficial for clinician-scientists involved in research endeavors.

## Reference Management Programs: Which Paper Had That Thing I Read That One Time?

One key method of staying organized during research projects is the use of a reference management program. Learning to use a reference management program can increase the speed of all aspects of a research project, including reviewing existing literature, composing institutional review board documents, and drafting and revising abstracts and manuscripts.<sup>6</sup> Some commonly-used reference management programs include: *EndNote* (Thomson Reuters, Philadelphia, PA, USA), *Mendeley* (Elsevier Inc., New York, NY, USA), *Zotero* (Roy Rosenzweig Center for History and New Media, Fairfax, VA, USA), and *RefWorks* (Proquest LLC, Ann Arbor, MI, USA). Most of these programs are available for free for end-users, either because they are open source, or because many universities and medical schools have an institutional

license. In addition, many manufacturers and academic libraries provide online or in-person training for these programs, which could increase the efficiency of use.

Key skills to acquire for the use of reference managers include: searching for references; importing references into the database; tagging references for future ease of use; searching for references in your database, especially searches based on keywords; and adding citations to your document (IRB/abstract/manuscript). Each of the above-mentioned reference management programs have their strengths and weaknesses;<sup>7</sup> choosing the program that will work best depends heavily on your preferences. Specifically, three areas in which the programs function differently include:

1. The workflow for importing documents into the database;
2. Methods available to query the reference database;
3. And whether the reference library is available and shareable online and on mobile devices.

Many comparisons of these software offerings exist, both in publications,<sup>7, 8</sup> and on the internet.

Developing an efficient method of importing references into a reference manager program will also help address what to do with all the papers that fellows and attending physicians tend to send out over email. Once added to the reference manager software, the papers will always be easily accessible and searchable.

One of the prime uses of a reference management program is inserting citations into text. Most of the reference manager programs have the capability to insert citations into word processing software (such as *Microsoft Word* (Microsoft, Redmond, WA, USA)) using journal-specific styles and with automatically updating numbering. In comparison to manually entering and updating citations, use of a reference management program saves significant amounts of time when documents are written, revised in response to reviewers, and resubmitted to another journal.

## Project Management Programs: What Was It I Was Supposed To Be Doing Today?

Project management programs can be useful in both the personal and professional realms. They are designed to help keep the various stages of a project organized, and can specifically assist in the process of

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planning the project, keeping track of progress, and adapting to changes in the project. Two examples of project management programs include: *Evernote* (Evernote Corp., Redwood City, CA, USA) and *Trello* (Trello, Inc., New York, NY, USA).

*Evernote* functions at its most basic level by making notes, which can contain text, checklists, and images. However, advanced versions of *Evernote* also have the ability to add and annotate PDFs, create notes with handwriting, store websites, and more (for a fee). *Trello* is based more on a whiteboard-style organization scheme, with boards representing projects and cards representing tasks for each project. This allows for granular task assignment to projects, and a different method of visualizing tasks for a given project. Both programs are accessible on most common devices (PCs, Macintosh, iOS, Android, etc.) and have slightly different interfaces depending on which device is being used. In addition, both programs allow sharing of tasks and notes to different users, allowing easy collaboration on projects.

The key functionality for any project management program is the near-real-time syncing capabilities across many devices. This way, portions of projects can be performed any time, in any place (with an internet connection), on any device. An inspiring idea about a new research project that occurs at the grocery store can be added to a note or card immediately, then later fleshed out, organized into a to-do list, and acted upon at a desk or remotely. This can minimize duplication of work, and help identify the next step of a project. Both of these programs can also be integrated into a digital calendar or have pop-up reminders; e.g. one can set a reminder for a month after paper submission to check on its progress, or remember the abstract deadline for the *American College of Cardiology Conference*. So far, the use of these programs in the medical setting has not been well-studied, though some examples exist.<sup>9</sup> There are numerous online tutorials for these programs, including some designed specifically for physicians.

#### Online File Storage: Did I Update the Right Version of That Draft?

Online file storage (otherwise known as “cloud storage”) is a method of storing files that frees the user from a specific computer or device. Online file storage programs usually have an automatic sync feature that continually updates files. So, if a document (e.g. manuscript draft) is edited on a work computer, and saved to the synced folder, the changes will then be reflected on the version of the file stored in the cloud, and then pushed to all other connected devices (e.g. smartphone, home computer). This allows easier transition of work from one computer to the next, allowing greater efficiency and a lower likelihood of editing the wrong version of a document. In addition,

online storage platforms can be used to share files between collaborators, even at different sites. Online storage does have potential limitations, though: not all systems are HIPAA-compliant, and not all are allowed by hospital/medical school IT departments. In addition, free accounts often have limited storage, and as with all computer systems, files can be accidentally deleted or damaged, so always back up important files.

#### Quick Hits: Bonus Efficiency Tips

In addition to the examples mentioned above, there are numerous other software tools designed to improve efficiency that could be useful for individual fellows and attending physicians, fellowship programs, and departments. For instance, some email services allow “snoozing” of emails, which then disappear until the snoozed time, decluttering the inbox. Some services provide shared calendars that any member can see and add events to, such as on call schedules and reminders for abstract and grant deadlines. Another example is voice recognition: some voice recognition programs can be integrated into electronic medical records, making writing clinic notes easier for those used to dictation.

#### Discussion

The utility of the approaches described above has not been studied in depth, but limited evidence suggests that health care professionals find using software tools useful.<sup>10</sup> The key is to find the set of tools that works best for any given individual for staying organized and working efficiently. Experimenting with these tools may take time up front, but could pay dividends later on; therefore, fellowship is an ideal time to develop a personal toolbox. As with any technology, development of new software tools might require a reevaluation of current preferences, but the potential benefit of that innovation is even more productivity.

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#### Biographical Sketch

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**Author Contributions:** AT conceived of and wrote the entire article.

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# Medical News, Products & Information

Compiled and Reviewed by Tony Carlson, Senior Editor

## Personalized Heart Models for Surgical Planning - System Converts MRI Scans into 3-D-Printed, Physical Models in a Few Hours

Researchers at MIT and Boston Children's Hospital have developed a system that can take MRI scans of a patient's heart and, in a matter of hours, convert them into a tangible, physical model that surgeons can use to plan surgery.

The models could provide a more intuitive way for surgeons to assess and prepare for the anatomical idiosyncrasies of individual patients. "Our collaborators are convinced that this will make a difference," says Polina Golland, a professor of Electrical Engineering and Computer Science at MIT, who led the project. "The phrase I heard is that 'surgeons see with their hands,' that the perception is in the touch."

This fall, seven cardiac surgeons at Boston Children's Hospital will participate in a study intended to evaluate the models' usefulness.

Golland and her colleagues will describe their new system at the International Conference on Medical Image Computing and Computer Assisted Intervention in October. Danielle Pace, an MIT graduate student in Electrical Engineering and Computer Science, is first author on the paper and spearheaded the development of the software that analyzes the MRI scans. Medhi Moghari, a physicist at Boston Children's Hospital, developed new procedures that increase the precision of MRI scans tenfold, and Andrew Powell, a cardiologist at the hospital, leads the project's clinical work.

The work was funded by both Boston Children's Hospital and by Harvard Catalyst, a consortium aimed at rapidly moving scientific innovation into the clinic.

MRI data consist of a series of cross sections of a three-dimensional object. Like a black-and-white photograph, each cross section has regions of dark and light, and the boundaries between those regions may indicate the edges of anatomical structures. Then again, they may not.

Determining the boundaries between distinct objects in an image is one of the central problems in computer vision, known as "image segmentation." But general-purpose image-segmentation algorithms aren't reliable enough to produce the very precise models that surgical planning requires.

Typically, the way to make an image-segmentation algorithm more precise is to augment it with a generic model of the object to be segmented. Human hearts, for instance, have chambers and blood vessels that are usually in roughly the same places relative to each other. That anatomical consistency could give a segmentation algorithm a way to weed out improbable conclusions about object boundaries.

The problem with that approach is that many of the cardiac patients at Boston Children's Hospital require surgery precisely because the anatomy of their hearts is irregular. Inferences from a generic model could obscure the very features that matter most to the surgeon.

In the past, researchers have produced printable models of the heart by manually indicating boundaries in MRI scans. But with the 200 or so cross sections in one of Moghari's high-precision scans, that process can take eight to 10 hours.



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## Interventional Pediatric Cardiologist

Levine Children's Hospital  
Sanger Heart & Vascular Institute  
Charlotte, NC

The Congenital Heart Center at **Levine Children's Hospital (LCH)** and **Sanger Heart & Vascular Institute (SHVI)**, seeks to add an additional Pediatric Interventional Cardiologist to join their existing faculty. Responsibilities for this position will include diagnostic and interventional catheterization, as well as general cardiology outpatient and inpatient care with shared night / weekend call.

- Candidates will have completed an ACGME accredited fellowship in pediatric cardiology and be BC/BE by the American Board of Pediatrics. A fourth year of additional training in interventional cardiac catheterization is required.
- A Minimum of 5 years of independent experience is preferred.

The Congenital Heart Center, established in 2010, has been ranked as one of the top-50 pediatric heart centers in the country by U.S. News and World Report for the last two years. Our comprehensive services include cardiac imaging, electrophysiology, dedicated cardiovascular intensive care staff, and regional referral programs in heart failure / transplantation, adult congenital heart disease, and fetal echocardiography. Surgical and cardiac catheterization volume has been growing at a rate of 15% per year over the last five years. Because of our continued growth, a new state of the art two lab cardiac catheterization and electrophysiology suite is under construction, with a plan to open in the fall of 2016. The cardiac catheterization program is active in industry sponsored clinical research.

LCH and SHVI are both premier referral facilities within the Carolinus HealthCare System (CHS), one of the nation's leading and most innovative healthcare systems. CHS operates nearly 2,500 system-employed physicians, more than 60,000 employees and more than 7,460 licensed beds across the Carolinas.

**Levine Children's Hospital (LCH)**, a state-of-the-art, 234 bed facility is the largest and most comprehensive children's hospital between Washington, DC and Atlanta, GA. LCH has a robust inpatient service with a 20-bed PICU/CVICU, 85-bed NICU, and an inpatient rehabilitation facility.

**Sanger Heart & Vascular Institute (SHVI)** is one of the largest cardiac and vascular programs in the Southeast, with more than 50 years of experience in providing world-class, comprehensive acute and chronic cardiovascular services, including the region's only heart transplant center and pediatric heart surgery program.

### To learn more or to submit your CV please contact:

Janisyn McLaurin, CHS Physician Recruiter at  
[Janisyn.mclaurin@carolinushealthcare.org](mailto:Janisyn.mclaurin@carolinushealthcare.org)

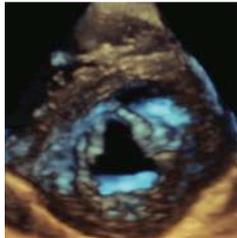
"They want to bring the kids in for scanning and spend probably a day or two doing planning of how exactly they're going to operate," Golland says. "If it takes another day just to process the images, it becomes unwieldy."

Pace and Golland's solution was to ask a human expert to identify boundaries in a few of the cross sections and allow algorithms to take over from there. Their strongest results came when they asked the expert to segment only a small patch --one-ninth of the total area -- of each cross section.

# Congenital and Paediatric Echocardiography: From Basics to Advanced

Thursday, April 28 to Saturday, April 30, 2016

The Hospital for Sick Children (SickKids)  
Daniels Hollywood Theatre, 1st Floor, Burton Wing  
555 University Avenue, Toronto, Ontario M5G 1X8

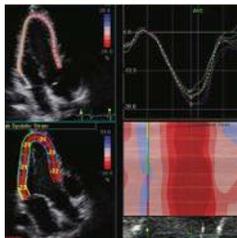
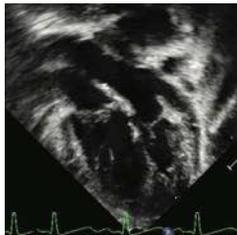


## Course Objectives

This course offers a unique introduction into Paediatric Echocardiography and imaging of Congenital Heart Disease. The main goal is to improve the knowledge of physicians and sonographers who perform and/or interpret echocardiograms in newborn and paediatric patient populations or perform echocardiographic studies in adults with repaired congenital heart disease.

Specific course objectives are:

1. Introduce the specific technical aspects of paediatric and congenital echocardiography: image acquisition and representation.
2. Introduction to cardiac morphology and how it relates to echocardiographic imaging.
3. Overview of pre-operative and post-operative imaging of the most common congenital defects.
4. Echocardiographic assessment of more complex congenital defects.
5. Introduction to assessment of systolic and diastolic function in children with a special focus on novel techniques for evaluating LV and RV function.



## Early Bird Registration (Before March 1, 2016)

Physicians . . . . .	\$495
Fellows/Sonographers/Trainees . . . . .	\$375

## Registration

Physicians . . . . .	\$595
Fellows/Sonographers/Trainees . . . . .	\$450

## Registration Fee Includes

- Attendance to all lectures
- Access to online course syllabus
- Continental breakfast, lunch and refreshments

## Accreditation

CME eligible

**SickKids**

[www.cvent.com/d/zfqjx4](http://www.cvent.com/d/zfqjx4)

"I think that if somebody told me that I could segment the whole heart from eight slices out of 200, I would not have believed them," Golland says. "It was a surprise to us."

Together, human segmentation of sample patches and the algorithmic generation of a digital, 3-D heart model takes about an hour. The 3-D-printing process takes a couple of hours more.

Currently, the algorithm examines patches of unsegmented cross sections and looks for similar features in the nearest segmented cross sections. But Golland believes that its performance might be improved if it also examined patches that ran obliquely across several cross sections. This and other variations on the algorithm are the subject of ongoing research.

The clinical study in the fall will involve MRIs from 10 patients who have already received treatment at Boston Children's Hospital. Each of seven surgeons will be given data on all 10 patients – some, probably, more than once. That data will include the raw MRI scans and, on a randomized basis, either a physical model or a computerized 3-D model, based, again at random, on either human segmentations or algorithmic segmentations.

Using that data, the surgeons will draw up surgical plans, which will be compared with documentation of the interventions that were performed on each of the patients. The hope is that the study will shed light on whether 3-D-printed physical models can actually improve surgical outcomes.

Guy Cloutier, also of the University of Montreal Hospital Research Centre, holds the promise of not only effectively identifying leaks, but also evaluating how the aneurysm is healing. And finally, the last word in endovascular repair is the bioactive stent and injection of a gel around the implant to prevent or stop leaks. These new biomaterials are being developed in collaboration with Soulez and Cloutier's colleague Sophie Lerouge.

Source: University of Montreal Hospital Research Centre (CRCHUM) @CRCHUMDr. Gilles Soulez is a radiologist with the University of Montreal Hospital (CHUM), a researcher at the University of Montreal Hospital Research Centre (CRCHUM), and a professor with the university's departments of Radiology, Radiation Oncology, and Nuclear Medicine. The University of Montreal is officially known as Université de Montréal.

## CONGENITAL CARDIOLOGY TODAY

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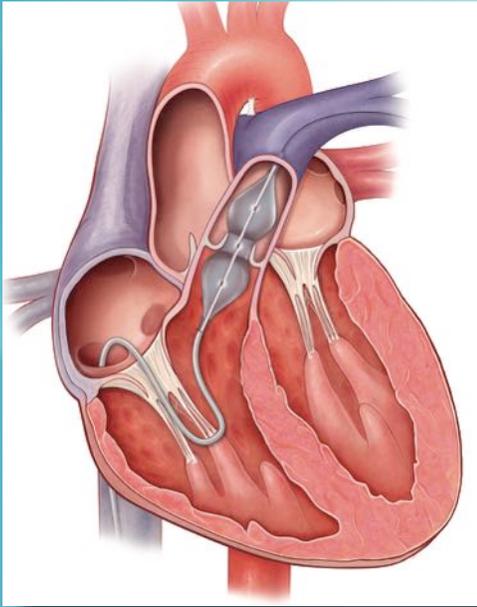
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