

# **Single-Stage Complete Repair Versus Multi-Stage Repair In Tetralogy Of Fallot With Borderline Pulmonary Arteries**

Thesis

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## *List of Abbreviations*

Abb.	Meaning
ACT.....	Activated clotting time
ASD.....	Atrial septal Defect
AV.....	Atrioventricular
AVSD.....	Atrioventricular Septal Defect
BT.....	Blalock-taussig
CHD.....	Congenital heart disease
CMR.....	Cardiac Magnetic Resonance
CT.....	Computed tomography
CTR.....	Cardiothoracic ratio
DescAo.....	Descending thoracic aorta
ECG.....	Electrocardiography
ECMO.....	Extracorporeal membrane oxygenation
ICU.....	Intensive Care Unit
IVC.....	Inferior Vena Cava
JET.....	Junctional Ectopic Tachycardia
LAD.....	Left Anterior Descending Artery
LPA.....	Left Pulmonary Artery
Lt.....	Left
LV.....	Left Ventricle
MBT.....	Modified Blalock-Taussig Shunt
MPA.....	Main Pulmonary Artery
MR.....	Magnetic resonance
NYHA.....	New York Heart Association
PA.....	Pulmonary Artery



## *List of Abbreviations*

<b>Abb.</b>	<b>Meaning</b>
PCO <sub>2</sub> .....	Partial carbon dioxide pressure
PDA.....	Patent Ductus Arteriosus
PGE <sub>1</sub> .....	Prostaglandin E <sub>1</sub>
PI.....	Pulmonary Incompetence or Insufficiency
PO <sub>2</sub> .....	Partial oxygen pressure
PVA.....	Pulmonary valve annulus
RBBB .....	Right Bundle Branch Block
RCA.....	Right Coronary Artery
RPA.....	Right Pulmonary Artery
Rt .....	Right
RV .....	Right Ventricle
RVOT .....	Right Ventricular Outflow Tract
RVOTO .....	Right Ventricular Outflow Tract Obstruction
SO <sub>2</sub> .....	Oxygen Saturation
SVC .....	Superior Vena Cava
SVT .....	Supraventricular Tachycardia
TAP .....	Transannular Patch
TTE .....	Transthoracic Echocardiography
VSD.....	Ventricular Septal Defect

## Abstract

### Objective:

Evaluation of the fate of patients having tetralogy of Fallot with borderline pulmonary arteries (McGoon ratio of 1.3 to 1.7) as regard to the need for a single-stage or multi-stage repair and the outcome of each surgical management.

### Patients and methods:

Between January 2016 and December 2017, 60 patients with tetralogy of Fallot and borderline pulmonary arteries were reviewed and divided randomly into 2 groups: •Group 1: 30 patients managed surgically by modified Blalock-Taussig (MBT) shunt as a part of multi-stage repair. •Group 2: 30 patients managed surgically by single-stage complete repair. The medical records of the patients were reviewed and age, sex, weight, and pre-operative oxygen saturation data were collected. All patients had pre-operative echocardiography and multi-slice computed tomography with angiography. Follow up was by echocardiography at discharge, 1 month and 6 months after operation. Multi-slice computed tomography with angiography was done in patients who had a shunt once the echocardiography showed acceptable pulmonary arteries.

### Results:

The patient age ranges from 5 to 50 months with a mean age of  $18.63 \pm 9.15$  ( $19.84 \pm 12.34$  for shunt group and  $17.43 \pm 8.54$  for repair group). The weight is ranging from 5kg to 18kg with mean of  $9.6 \pm 2.53$  ( $8.82 \pm 2.79$  for shunt group and  $10.41 \pm 2.63$  for repair group). The mean pre-operative O<sub>2</sub> saturation was  $68.95\% \pm 7.8\%$  for shunt group and  $87.93\% \pm 6.18\%$  for repair group. The median McGoon ratio for shunt group is 1.4 and for repair group is 1.6 with a highly significant statistical difference ( $p < 0.0001$ ). Mortality rate in our study is 10% (10% for shunt group and 10% for repair group). Morbidity incidence rate is (26.6%) for shunt or repair group. The ICU stay ranges from 2 to 31 days with a median 3 days for the shunt group (mean of  $3.61 \pm 1.91$ ), and median 4 days for the repair group (mean of  $6.07 \pm 6.63$  days). Calculated p value showed significant difference between the 2 groups as regard to ICU stay. The post-operative SO<sub>2</sub> significantly rose to mean of  $85.58 \pm 7.05$  in the shunt group and to  $98.14 \pm 3.36$  in the repair group ( $p < 0.00001$ ).

### Conclusion:

There is no great difference between multi-stage repair and single stage complete repair regarding to morbidity and mortality but regarding to ICU stay the single-stage is longer. McGoon ratio of 1.5 can be used as a guide point in the management decision making.

# INTRODUCTION

**T**etralogy of Fallot (TOF) is a congenital cardiac malformation consists of ventricular septal defect, aortic overriding, infundibular pulmonary obstruction, and right ventricular hypertrophy (*Anderson et al., 1981*).

Tetralogy of Fallot is the commonest cyanotic heart disease with prevalence from 0.21 to 0.36 per 1000 livebirths, and reaching about 7 to 10 % of all forms of congenital heart diseases (*Ferencz et al., 1985 and Mitchell et al., 1971*).

Tetralogy of Fallot had been uniformly lethal, but this changed after the introduction of the systemic to pulmonary shunt by Alfred Blalock (*Taussig et al., 1947*).

On November 29, 1944, Alfred Blalock performed the 1<sup>st</sup> operation on a cyanotic 1 year-old child with TOF. The Blalock-Taussig (BT) shunt had a high rate of success, and by 1950, 1000 such operations had been performed by Blalock and his team (*Taussig et al., 1979*).

The 1<sup>st</sup> intracardiac repair of TOF was by C. Walton Lillehei and his team at the University of Minnesota (*Warden et al., 1954*).

Surgical management of TOF is still a debatable subject, especially whether to perform single stage complete repair or to perform systemic to pulmonary shunt surgery and delaying the

complete repair. When and to which patients each surgical strategy should be applied is a very complicated subject (*Stewart et al., 2013*).

Mortality rate in cases managed by single-stage complete repair has decreased a lot. From 10 cases performed by Dr. Lillehei 6 patients only survived (*Lillehei et al., 1955*). Mortality was 0.5% among 366 patients between 1980 and 1991 reported by Roger Mee and his colleagues (*Karl et al., 1992*) , and it decreased up to 0% in a series of 102 patients with average age 5.9 months at Children's Memorial Hospital in Chicago between 1997 and 2004 (*Stewart et al., 2005*).

Even so, some centers still perform systemic to pulmonary shunt especially modified Blalock-Taussig (MBT) shunt in the neonatal period followed by complete repair later on (*Morales et al., 2009*).

Recent studies showed equivalent survival between the two strategies (*Kanter et al., 2010*).

Shunting or single-stage repair of neonates with symptomatic TOF provides equivalent mortality and results. Shunted patients had fewer transannular patch repairs despite having more emergent initial operations. Compared with the single-stage repair group, shunted patients had decreased intensive care unit and hospital stays for the 1st hospitalization,

which were neutralized when the second operation (repair) values were added (*Kanter et al., 2010*).

The size of the pulmonary arteries is very important prognostic factor in TOF management because if they are too small may lead to right ventricular (RV) failure after separation of pulmonary and systemic circulations if the outflow resistance remains too high (*Laas et al., 1984 and Harlan et al., 1995*).

## **AIM OF THE WORK**

**T**his study aims at evaluating the fate of 60 patients having tetralogy of Fallot with borderline pulmonary arteries (McGoon ratio of 1.3 to 1.7) as regard to the need for a single-stage or multi-stage repair and the outcome of each surgical management.

## Chapter 1

# **ANATOMY OF TETRALOGY OF FALLOT**

**T**here is four subgroups of TOF exist:

- 1- Tetralogy of Fallot with pulmonary stenosis.
- 2- Tetralogy of Fallot with absent pulmonary valve syndrome.
- 3- Tetralogy of Fallot with common atrioventricular canal (AVSD).
- 4- Tetralogy of Fallot with pulmonary atresia which is perhaps best characterized as being at the extreme end of tetralogy of Fallot and differs from regular tetralogy of Fallot in having obligatory extracardiac sources of pulmonary blood flow

*(Stewart et al., 2013)*

In this study tetralogy of Fallot with pulmonary stenosis will be particularly discussed.

It is consists of 4 major components which vary in its details, these 4 are:

- 1- Right ventricular outflow tract obstruction (RVOTO).
- 2- Overriding aorta.