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Early Outcomes of Pulmonary Valve Replacement After Total Correction of Tetralogy of Fallot

Thesis

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ABSTRACT

Introduction: Severe right heart failure with serious consequences, may develop after pulmonary regurgitation (PR) caused by total correction of tetralogy of Fallot (TOF). This thesis is made to present early outcomes of surgical Pulmonary valve replacement (SPVR) in these patients.

Methods: From 2019-2021, 38 patients 25 male 13 female mean age was 15.76 ± 6.24 years underwent SPVR after 12.63 ± 4.76 years from first operation. Patients underwent for SPVR with either tissue or mechanical valves using cross clamp and giving cardioplegia. Repair of tricuspid, closure of residual ventricular defect (VSD), resection of residual pulmonary stenosis (PS) or right ventricle (RV) aneurysms resection may be indicated during procedure. Follow up X-ray and transthoracic echocardiography (TTE) made after 6 months and one year after procedure. Follow up cardiac magnetic resonance imaging (CMRI) made on average from one to three years after procedure.

Results: there was no perioperative mortality. Mean of hospital stay was 6.74 ± 1.08 days. the results of follow up x-ray, TTE and CMRI demonstrate that after SPVR the RV experiences improvement on its volumes and systolic function.

Conclusion: SPVR seems to be a positive approach and recommended to be done in correct timing.

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List of Abbreviations

Abbr.	Full term
2D	Two-dimensional
3D	Three-dimensional
3DR	Three-dimensional reconstruction
ASD	Atrial septal defects
BMI	body mass index
CHD	congenital heart disease
CMRI	Cardiac magnetic resonance imaging
CIED	cardiac implantable electronic device
CTR	Cardiothoracic ratio
CRT	cardiac resynchronization therapy
EDFF	End-diastolic forward flow
EF	Ejection fraction
HPVR	Hybrid pulmonary valve replacement
ICDs	implantable cardioverter defibrillators
IVC	Inferior vena cava
IUD	Intrauterine device
LAD	Left anterior descending
LV	Left ventricle
LVEDVI	Left ventricular end diastolic volume indexed
NSF	nephrogenic systemic fibrosis

NT-proBNP	N-terminal pro brain natriuretic peptide
NYHA	New York Heart Association
PG	Pressure gradient
PR	Pulmonary regurgitation
PRF	Pulmonary valve regurgitation Fraction
PV	Pulmonary valve
PVR	Pulmonary valve replacement
RV	Right ventricle
RVEDVI	Right ventricular end diastolic volume indexed
RVEF	Right ventricular ejection fraction
RVESVI	Right ventricular end systolic volume indexed
RVOT	Right ventricular outflow tract
SPVR	Surgical pulmonary valve replacement
TAP	Transanular patch
TAPSE	Tricusped annular plane systolic excursion
TOF	Tetralogy of fallot
TPVR	Transcatheter pulmonary valve replacement
TR	Tricuspid regurgitation
TTE	Transthoracic echocardiographic
VSD	Ventricular Septal defect
VT	ventricular tachycardia

Introduction

Tetralogy of Fallot (TOF) is the most common form of cyanotic congenital heart disease (CHD) at birth and occurs at a rate of 0.28–0.48 per 1000 live births. It consists of Right ventricular outflow tract (RVOT) obstruction, Ventricular Septal defect (VSD), Right ventricular (RV) hypertrophy and overriding of aorta **[1]**.

It also continues to be the most common cyanotic congenital heart defect with survival into adulthood. Survival is now expected into adulthood, and accounts for 3.5%–10% of all congenital defects **[2]**.

Surgical repair of TOF includes closure of the VSD, and relief of RVOT obstruction that involves RVOT infundibular muscle resection, pulmonary valvotomy or valvectomy, and commonly RV outflow augmentation with placement of a subvalvular or transannular patch **[3]**.

Despite the profound impact of surgical intervention on functional status, survival, and quality of life, **[4]** postoperative residua and sequelae are expected in patients with repaired TOF and lifelong informed follow-up is required **[6]**.

Health care providers across the world are trying to take care of the complications that may be seen years after the repair. In TOF complications include: pulmonary valve regurgitation, heart failure, ventricular tachycardia, atrial tachyarrhythmias, and sudden cardiac death **[5]**.

Approximately 50%–60% of TOF patients will die from a cardiac related death; of these cardiac related deaths, sudden cardiac death (17%–36%) and heart failure (14%–24%) are the most common. Another very important complication is pulmonary regurgitation (PR) that will be present in up to 50% of TOF patients, of whom 37% will need reoperation **[6]**.

PR occurs with old routine techniques of surgical repair of TOF like: extensive ventriculotomy, and infundibulectomy as well as generous transannular patching of the RV outflow tract **[7]**.

TOF patients are the most group in adulthood whom undergo for reoperation, 37% of reoperations were pulmonary valve replacements (PVR), and the timing for it is a topic of much interest since earlier operation might prevent deleterious effects later in life **[5]**.

PVR may be indicated for the purpose of preventing permanent alterations of cardiac walls and hemodynamics. Although PVR performed by numerous surgeons for many years, the effect of PVR still remains controversial. Despite the evidences of recovered cardiac function and hemodynamics post-PVR, the beneficial effect of PVR on prevention of alterations of ventricular mass, volumes, and hemodynamics remains unclear and indications for PVR are restricted to limited conditions such as RV dysfunction **[8]**.

Aim of Work

The aim of this thesis is to study the early outcomes of SPVR after previous total correction of TOF with the duration of follow up ranged from one to three years after operation.

Anatomy & physiology of RV & pulmonary valve

The pulmonary valve is the most anterior valve and differs from the aortic valve in that it lacks continuity with the atrioventricular valves. It rises from the ventricular septum through the subpulmonic infundibulum or conus. The conus is a remnant of the bulbus cordis that disappears on the left ventricle (LV) outflow and shortens in the RV outflow. An anterior displacement of the conus is responsible for the subpulmonary stenosis in TOF **[9]**.

The RV is crescent shaped and wraps around the LV. In normal hearts, the RV has only two layers, whereas the LV has myofibers arranged in three different orientations (oblique in the surface, longitudinal on the subendocardium, and circumferential in between the two layers). In histologic studies, patients with TOF were found to have a midlayer formed of circumferential myofibers **[10]**.

The two ventricles besides sharing spiraling muscle bundles, share an interventricular septum and pericardium. All these anatomical landmarks are important since they are the basis of the "interventricular dependence" and explain