DECLARATION

I, Deliwe Precious Ngwezi declare that this research report is my own work. It is being submitted for the degree of Master of Medicine in the branch of Paediatrics in the University of the Witwatersrand, Johannesburg. It has not been submitted before for any degree or examination at this or any other University.

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ABSTRACT

BACKGROUND: Despite the advances in the surgical treatment of Tetralogy of Fallot(TOF), pulmonary regurgitation (PR) remains as a possible complication with detrimental impact on right ventricular function (RV) in the long term. If these effects are not reversed timeously by replacing the pulmonary valve, serious morbidity and even mortality may occur.

AIM: The aim of the study was to assess the outcomes in children who had complete repair of TOF at Charlotte Maxeke Johannesburg Academic Hospital (CMJAH) with or without a RV outflow conduit with emphasis on the development of PR, as well as signs and symptoms that could be linked to PR.

METHOD: A retrospective clinical audit on patients with a diagnosis of TOF and who had surgery at the CMJAH between 01/01/1994 and 31/12/2003 was undertaken. Data collected and entered onto a data collection sheet included ethnicity, sex, type of surgery which was either palliation with a Blalock-Taussig shunt (BTS) or corrective surgery, age at surgery, echocardiographic documentation of PR and RV size during follow-up. Other information included electrocardiogram (ECG) changes, abnormalities detected on chest X-ray (CXR), whether or not the patient was subjected to re-operation, as well as the indications of re-operation.

RESULTS: Of the 72 patients with TOF and available data, fifty four patients (75%) had surgery, 50 (92.6%) of whom had corrective surgery and 4(7.4%) had palliative surgery in the form of a BTS only. Of the 50 who had corrective surgery, 2 (4%) patients had an initial BTS shunt while the remaining forty eight (96%) underwent initial primary corrective surgery. There were 46 males (63.9%) and 26 (36.1%) females. The median age at first visit was 13 months (range, 0.03 to 177), median age at corrective repair was 39.5 months (range, 3 to 210) and median age at palliation was 18 months (range, 1.5 to 29). Of the 50 patients undergoing corrective surgery, 60% had complex anatomy and 40% simple anatomy. More females compared to males had complex anatomy with a hypoplastic pulmonary valve (p= 0.047). Two patients undergoing a transannular patch (TAP) developed severe PR in the immediate postoperative period (less than 1 year) whilst no patients in the simple repair group developed severe PR (p= 0.110). In the intermediate postoperative period (1 to 5 years), there was 1 patient each in the TAP and simple repair group with severe PR (p = 0.476). Finally in the long term period (more than 5 years), 8 patients with TAP developed severe PR whilst 2 patients with simple repair developed severe PR (p= 0.005). Two patients out of fifteen patients (13.3%) with TAP had a pulmonary valve replacement (PVR). No patients with simple repair had PVR.

CONCLUSION: Severe PR is an expected and serious complication accompanying all forms of repair techniques employed for the wide anatomical spectrum of TOF. In this study, comparison of the two most common types of repair, namely simple versus TAP, revealed a statistically significant occurrence of PR in the TAP group. Appropriate timing for PVR is paramount for the symptomatic patients in order to preserve RV function.

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TABLE OF CONTENT **PAGE**

LIST OF TABLES

LIST OF FIGURES

ABBREVIATIONS

BTS	Blalock-Taussig Shunt
BWIS	Baltimore Washington Infant Study
CHD	Congenital Heart Disease
CMJAH	Charlotte Maxeke Johannesburg Hospital
CMRI	Cardiac Magnetic Resonance Imaging
CP	Cerebral Palsy
CXR	Chest x-ray
ECG	Electrocardiogram
ECHO	Echocardiogram
MR	Muscle Resection
NYHA	New York Heart Association
PA	Pulmonary Atresia
PR	Pulmonary Regurgitation
PS	Pulmonary Stenosis
PTFE	Polytetrafluorethylene
PV	Pulmonary Valve
PVR	Pulmonary Valve Replacement
RAD	Right Axis Deviation
RAE	Right Atrial Enlargement
RBBB	Right Bundle Branch Block
RPA	Right Pulmonary Artery
RVH	Right Ventricular Hypertrophy
RV	Right Ventricle

RVOT	Right Ventricular Outflow Tract
TR	Tricuspid Regurgitation
TOF	Tetralogy of Fallot
TAP	Transannular Patch
VCFS	Velocardiofacial Syndrome
VE	Ventricular Ectopics
VSD	Ventricular Septal Defect