T♥F-LIFE
Tetralogy of Fallot for Life

# A Prospective Multi-Centre International Cohort Study

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Population Health Research Institute
HEALTH THROUGH KNOWLEDGE

Hamilton Health Sciences

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HEALTH SCIENCES

Background

- 38,000 patients diagnosed annually

- Multiple surgical strategies

- Late RV dysfunction and mortality

Central Question: Does surgical repair technique impact on RV health and survival in anatomically equivalent TOF patients?

Hypothesis

Equivalent Anatomy

V
S

**Right Ventricular Physiology & Morphology**
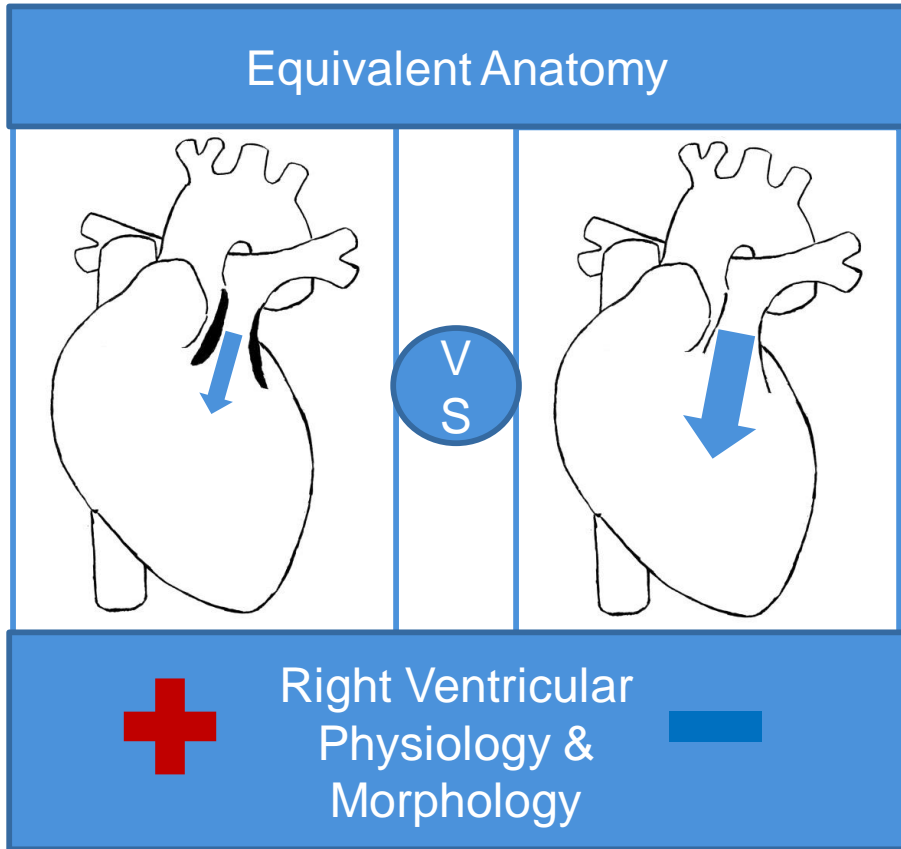
For equivalent preoperative anatomy, those having little to no right ventricle outflow obstruction and free pulmonary regurgitation will have more right ventricle dilation than those having mild to moderate residual right ventricular outflow tract obstruction and more limited pulmonary regurgitation at 2 years following repair

Design

- Prospective observational cohort
- 1100 participants
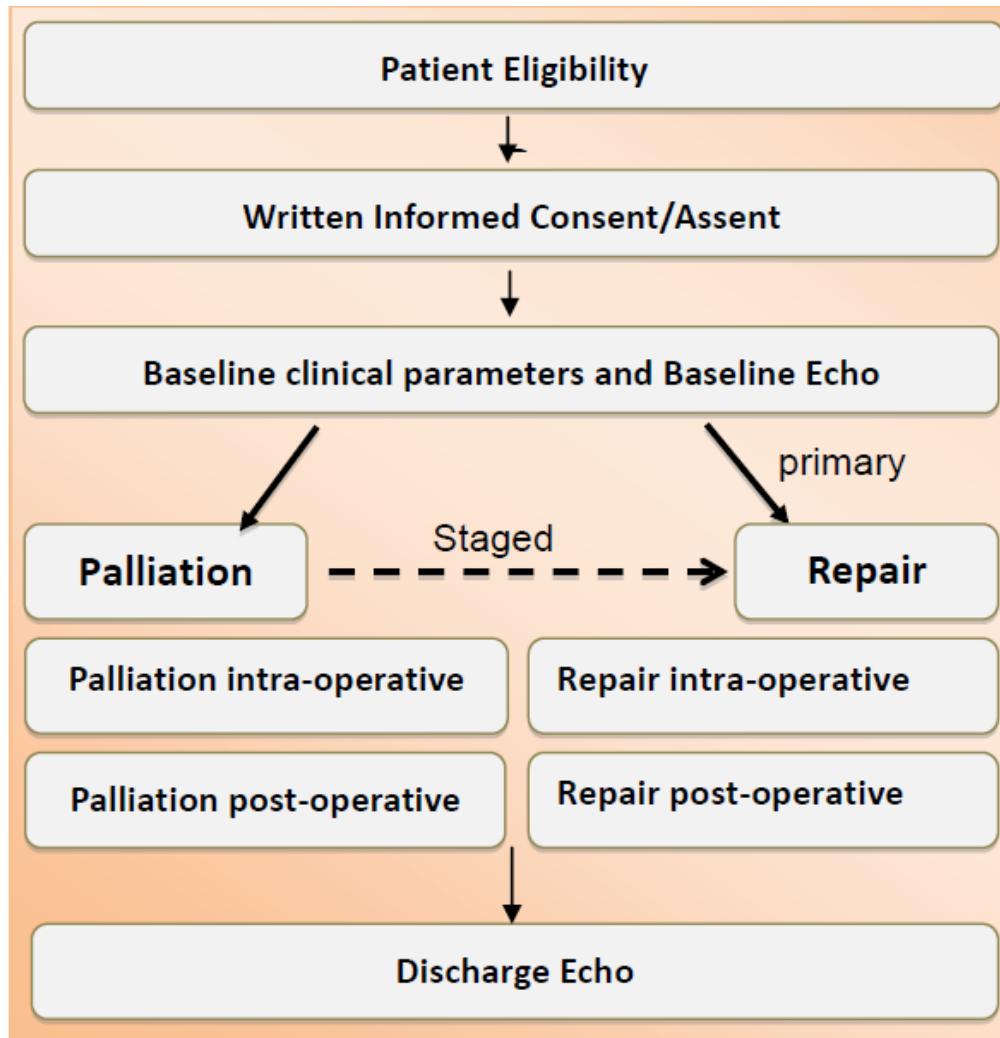
# Follow-up
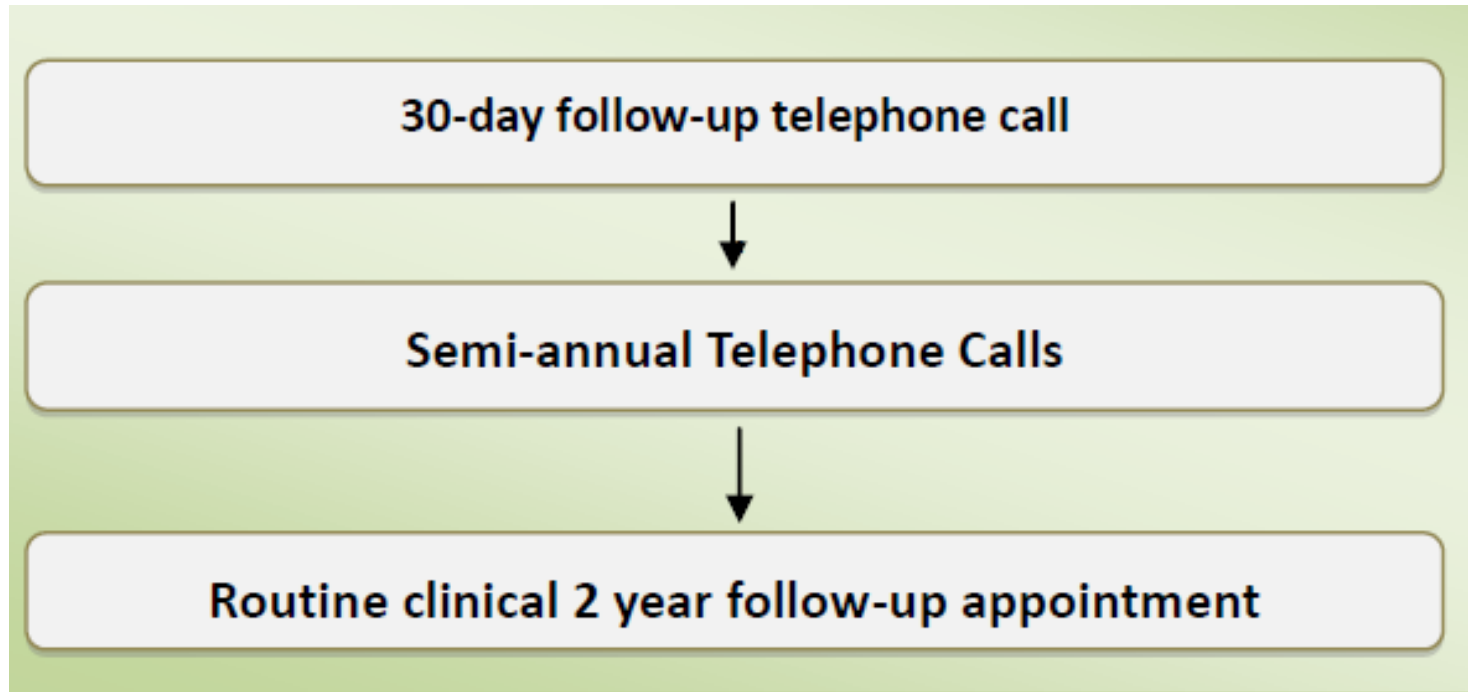
Eligibility

Inclusion Criteria

- Patient presenting with tetralogy of fallot (TOF) with right ventricular outflow tract (RVOT) or pulmonary valve stenosis or pulmonary atresia and confluent pulmonary arteries (no MAPCA's)

- Admitted with an intent to treat (i.e. patient planned to undergo a primary or staged repair)

Eligibility (cont'd)

Exclusion Criteria

- TOF with absent pulmonary valve

- Other major cardiac anomalies such as AVSD, multiple VSDs, right atrial isomerism and major aortopulmonary collateral arteries

- Unbalanced ventricles precluding biventricular repair

- Major genetic abnormalities/syndromes (e.g. trisomy 13,18, and 21)

- Major non-cardiac anomalies (e.g. diaphragmatic hernia, omphalocele, absent sternum)

- Infective endocarditis prior to cardiac intervention (palliation or repair)

- Stroke in the last 30 days prior to cardiac intervention

- Known diagnosis of HIV or hepatitis B

- Any previous cardiac procedures

Primary Objective
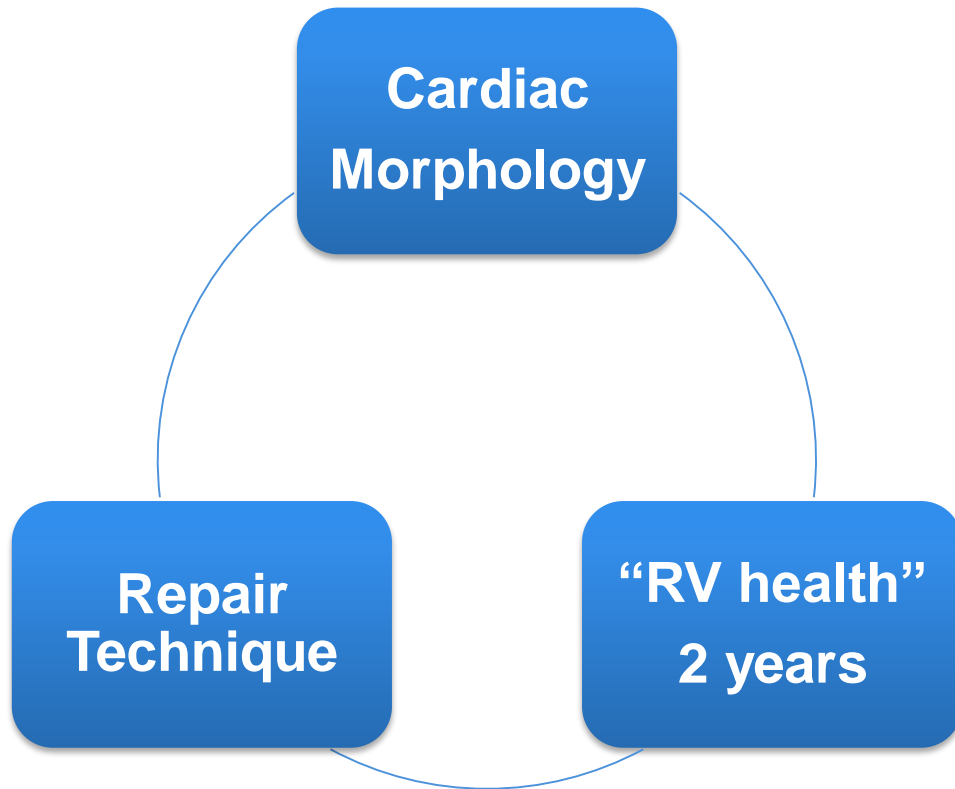
Endpoints (echo at 2 years):
- Indexed RV diastolic dimension
- RV/LV diastolic diameter ratio
- RV and LV function
- Residual RVOT obstruction
- Pulmonary insufficiency

Secondary Objectives

To determine:

- The pattern of palliation procedures (BT shunt, RVOT stent, or balloon dilation), surgical repair strategy (staged versus primary repair), and surgical repair technique (RVOT management: AP, minimal TAP, and standard TAP; Approach to VSD closure: trans-atrial, trans-ventricular) at participating centres

- The 30-day and 2 year cardiovascular mortality rate (for equivalent patients) after primary and staged repair

- The 30-day and 2 year cardiovascular mortality rate (for equivalent patients) after various surgical repair techniques for VSD closure and RVOT management

- The rate of palliation failure following various palliation techniques

- The interval mortality rate following various palliation techniques (BT shunt, RVOT stent, balloon dilation)

- The **possible** effect of palliative procedures (BT shunts, balloon dilation, stent insertion) on cardiac morphology (growth of the infundibular chamber, the pulmonary annulus and PA branches' diameter) and subsequent repair technique

Secondary Objectives (cont'd)

- The relationship between repair technique/strategy and prevalence of post-operative restrictive physiology as defined by the presence of antegrade flow in pulmonary artery during atrial contraction on echocardiogram

- The association between the repair strategy (primary or staged repair) with RV physiology and morphology at 2 years (for equivalent patients)

- The right ventricular morphological and physiological adaptations to severe pulmonary stenosis or regurgitation using repaired TOF pulmonary atresia as a model

- The relationship between TOF repair strategy/technique on the incidence and prevalence of cardiac re-interventions (e.g. pulmonary valve implantation, RVOT stent insertion or balloon dilatation)