La Maladie Bleue- Surgical Success of Tetralogy of Fallot

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Introduction

Background

- Louis Arthur Etienne Fallot first reported 'La Maladie Bleue' in 1888- this is the clinical term for the physiology caused by a collection of anatomical abnormalities today known as Tetralogy of Fallot (ToF).
- ToF is a congenital heart condition consisting of four heart defects: ventricular septal defect (VSD), a hole in the septum between the left and right ventricles of the heart- caused by a deviation in the developing ventricle and leading to the combining of oxygenated and deoxygenated blood; pulmonary stenosis, narrowing of the pulmonary valve; an overriding aorta, one which has shifted towards the VSD in order to compensate for the reduced blood flow; and ventricular hypertrophy, the thickening of the right ventricular wall.
- It is also the most common cyanotic heart condition in children who have survived beyond neonatal age without treatment.
- ToF's surgical approaches reflect advancement in cardiology, cardiac surgery, and knowledge of the developing heart.

Aims of study

- To investigate the types of surgical corrections on ToF.
- To evaluate the success of surgical corrections on ToF patients.
- To investigate future treatment for ToF.

Methods

- There are two types of repair: complete and temporary.

Complete Repair

- 2 main procedures occur: a transannular patch (TAP) directly over the VSD, or a pulmonary valve-sparing procedure (PVS) to widen the pulmonary valve.
- Despite this surgery, the valve may still be too narrow, and the patient may require replacement valves.
- Furthermore, stents can be used to increase blood flow in the right ventricular outflow tract.

Temporary Repair

- Shunts are a type of palliative surgery on infants used in extreme cases of ToF.
- The development of the Blalock-Thomas-Taussig shunt surgery was a ground-breaking discovery. Taussig found that children with a cyanotic heart defect and a patent ductus arteriosus (PDA- a medical condition in which the ductus arteriosus fetal blood vessel fails to close) lived longer than those without the PDA. Her shunt performed similarly to a PDA and improved the oxygenation of individuals with ToF.

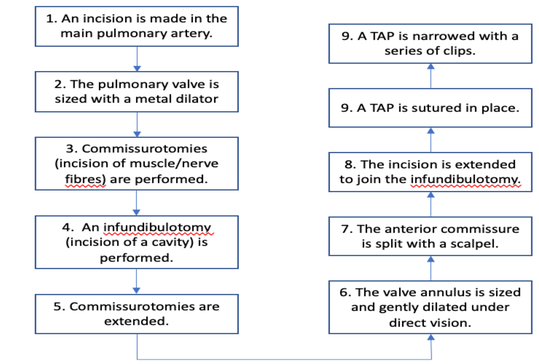

Figure 1. flow chart of TAP procedure

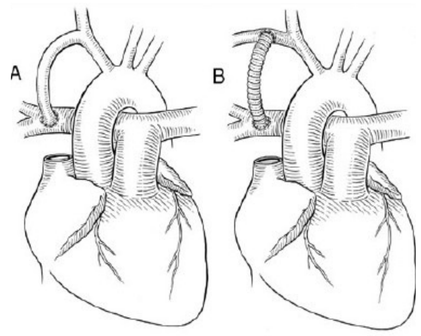

Figure 2. Blalock–Thomas–Taussig shunt A- original procedure B- modified procedure (Pezard et al., 2023)

Results

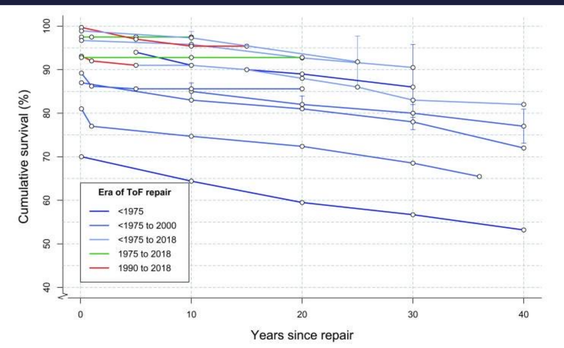

Figure 3. Line chart exhibiting Survival following tetralogy of Fallot (ToF) repair. Dots reflect Kaplan-Meier survival estimates at various time points, whereas coloured lines denote a particular study. Where published, vertical lines are used to depict 95% confidence intervals. According to the surgery era, lines are coloured (van der Ven et al., 2019).

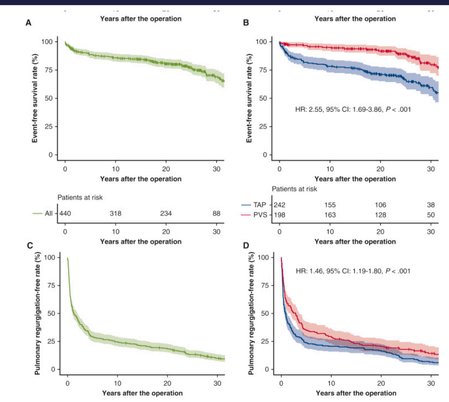

Figure 4. Overall survival in all study cohorts (A) and by procedures (B). Adverse event-free survival in all cohorts (C) and by procedures (D. Bands above and below the fitted line represented 95% CIs. HR, Hazard ratio; CI, confidence interval; TAP, transannular patch; PVS, pulmonary valve sparing.(Ono et al., 2022)

Discussion

Discussion

- The main goal for ToF treatment is to repair the VSD with open heart surgery at birth or during infancy: 1 year or younger.
- In the past, TAP repair procedures were more common. However, the enlargement of the right ventricle from progressive pulmonary insufficiency has been observed.
- In recent years and for the foreseeable future, there has been a trend towards protecting the right ventricle's function through maintaining the pulmonary valve's functionality. Recent research shows that the PVS method had tolerable pulmonary stenosis and, in well-selected patients, considerably reduced the frequency of moderate to severe pulmonary regurgitation and other related problems of TAP repair.
- PVS is the best surgical technique for repairing TOF completely, however it's debated how to distinguish between individuals who can have PVS and those who need TAP.
- Tetralogy of Fallot patients require lifetime monitoring and additional treatments (including ECGs, echocardiography and exercise testing). Even as adults, additional treatments (such stenting for pulmonary stenosis) could be necessary.
- However, the surgical repair of ToF has led to significant increase in mortality and long-term survival is almost 90%

Limitations

- Limitations for the study producing Figure 3 include lack of understanding of right ventricular (RV) adaptation and failure and how it might affect mortality rate despite the modelling completed, and indications differing from guidelines such as the 2009 CCS guideline which do not take into account the RV volume differences between genders and ages.
- Limitations for the study producing Figure 4 include inability to control minor events that might skew results, Z score of pulmonary valve diameter in PVS patients rarely being <-2.0, age of cohort examined being slightly older than usual operative age.

Conclusion

- The mortality risk associated with total repair of tetralogy of Fallot has slowly decreased over time. The care of ToF patients should be based on the severity of pulmonary stenosis, recognition of later complications, and changes in modern medicine. Although ToF patients will receive lifelong healthcare, with the right treatment and attention early on, patients are able to lead a normal, healthy life

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