

Surgical options for tetralogy of Fallot

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- Anatomy
- Physiology

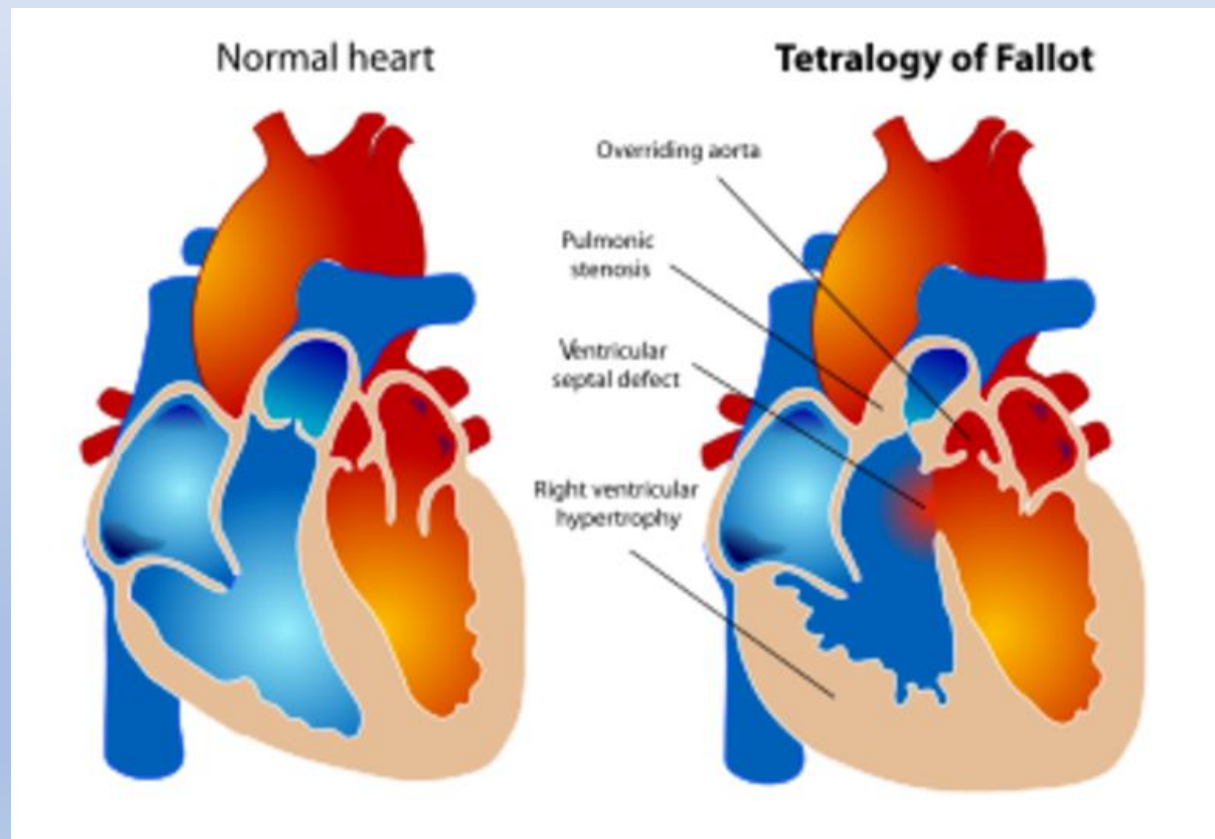
- Children
- Adults
- Complications
- Follow up



Anatomy

Etienne Fallot (1850-1911)

- VSD
- Overriding aorta
- Pulmonary stenosis
- Hypertrophied RV
- + ASD = pentalogy of Fallot (!)



- **VSD** (hole in the heart)
- **PS** (obstruction in the passage of blood to the lungs)
 - Small annulus / bad leaflets
 - Multilevel

The modern anatomic description is 'antero-cephalad deviation of the outlet septum'.

The aorta and RVH are byproducts of the anatomy and the physiology.

Physiology, presentation

ToF is a spectrum of disease

The **pulmonary valve** determines the presentation and the management

Pulmonary obstruction can be **multilevel** – RVOT, PS, PA

Physiology, presentation clinical spectrum

- Very little PS – the physiology is that of a large VSD - L-R shunt, L heart overload, saturations of 100% (pink Fallot) and potentially heart failure
- Severe PS – not enough blood going to the lungs → blue baby

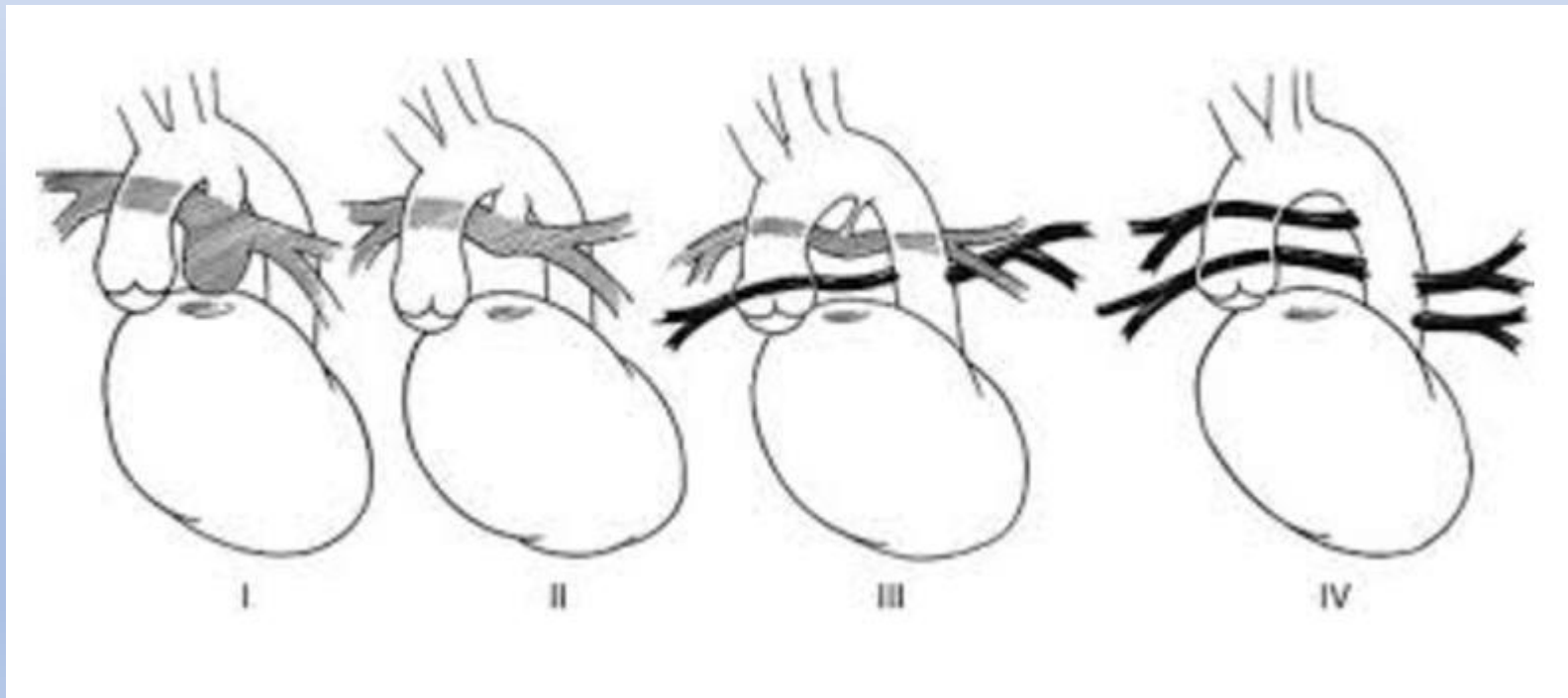
The PV may be adequate initially but when the RV hypertrophies the multilevel obstruction gets worse → cyanotic spells (hyperacute R-L shunt)

Variants of ToF based on the pulmonary valve

- ToF and pulmonary **stenosis** – most common
- ToF and pulmonary **atresia** – there is no communication between RV and main pulmonary artery (PA/VSD), a duct-dependent lesion
- ToF and **absent** pulmonary valve – there is wide communication with the pulmonary artery but not guarded by a valve (absent!) hence free PR

ToF and pulmonary atresia (PA/VSD)

..... SPECTRUM



ToF and absent pulmonary valve

- Giant PAs
- Airway obstruction

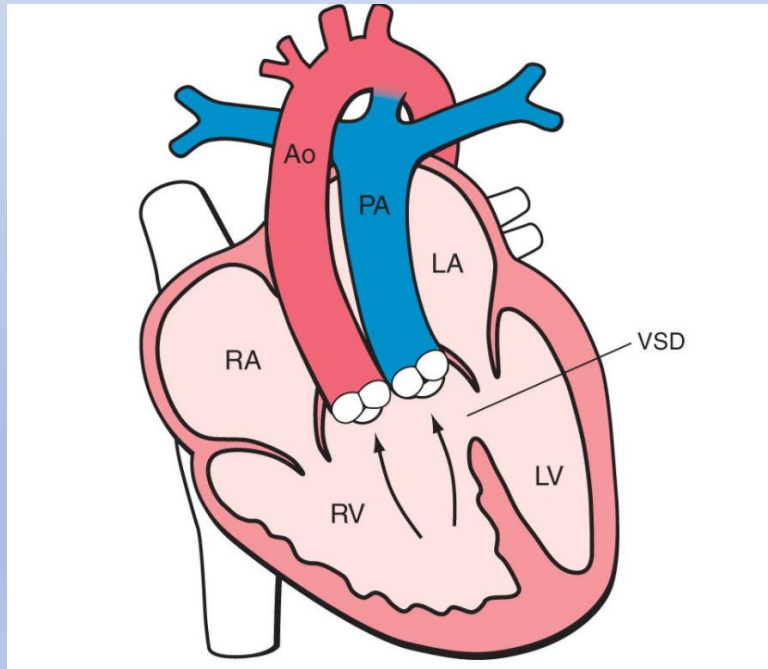
At operation the PAs have to be made smaller rather than bigger (!)



DORV = double outlet right ventricle

- more than 50% Ao override

- a **spectrum** between ToF and transposition



Anatomy



Physiology



Management

Average patient = 6-month old with occasional spells

Problem = not enough blood going to the lungs

Solution = bring more blood to the lungs

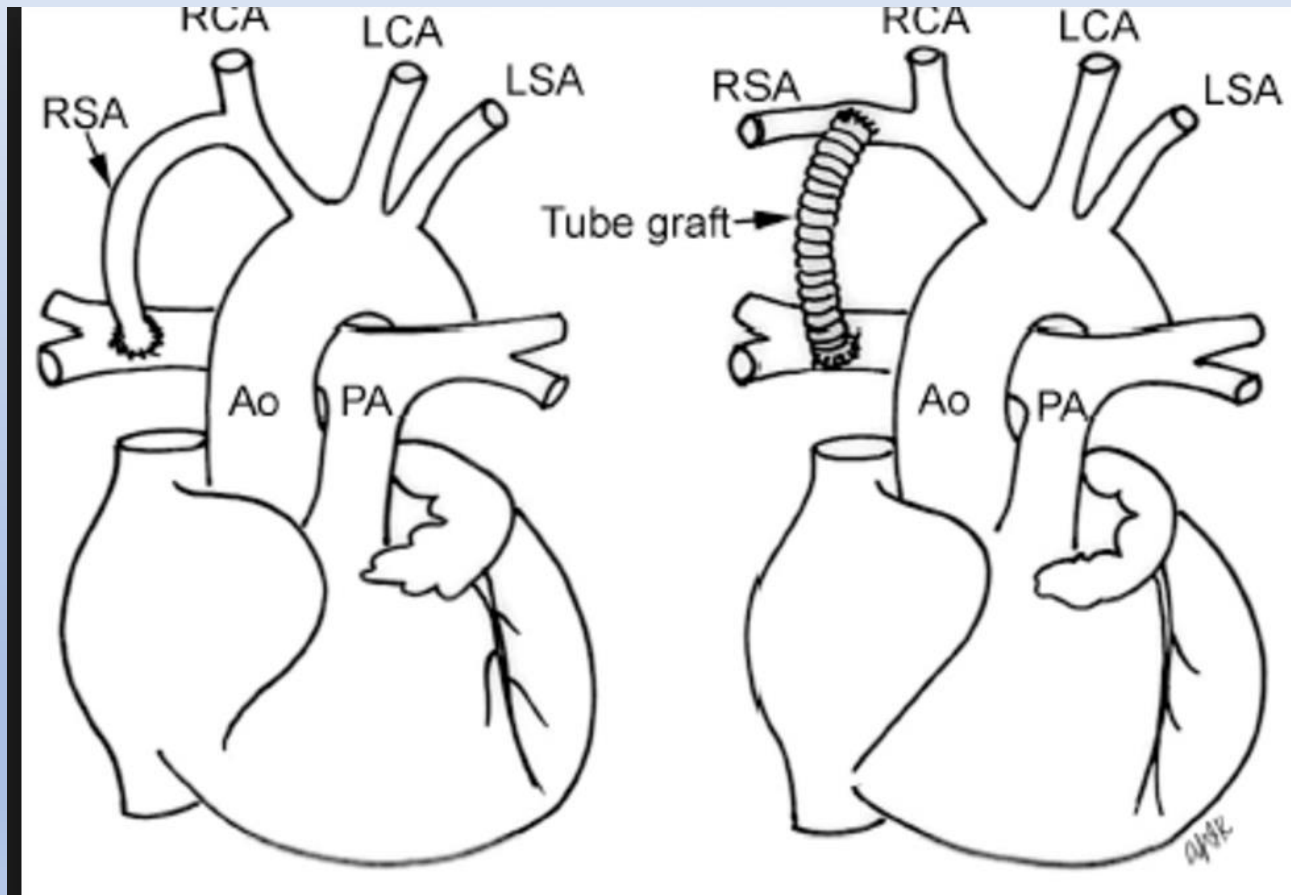
Historically – Blalock-Taussig shunt

A form of palliative treatment which helps the patient to continue growing until an operation can be done

BT shunt

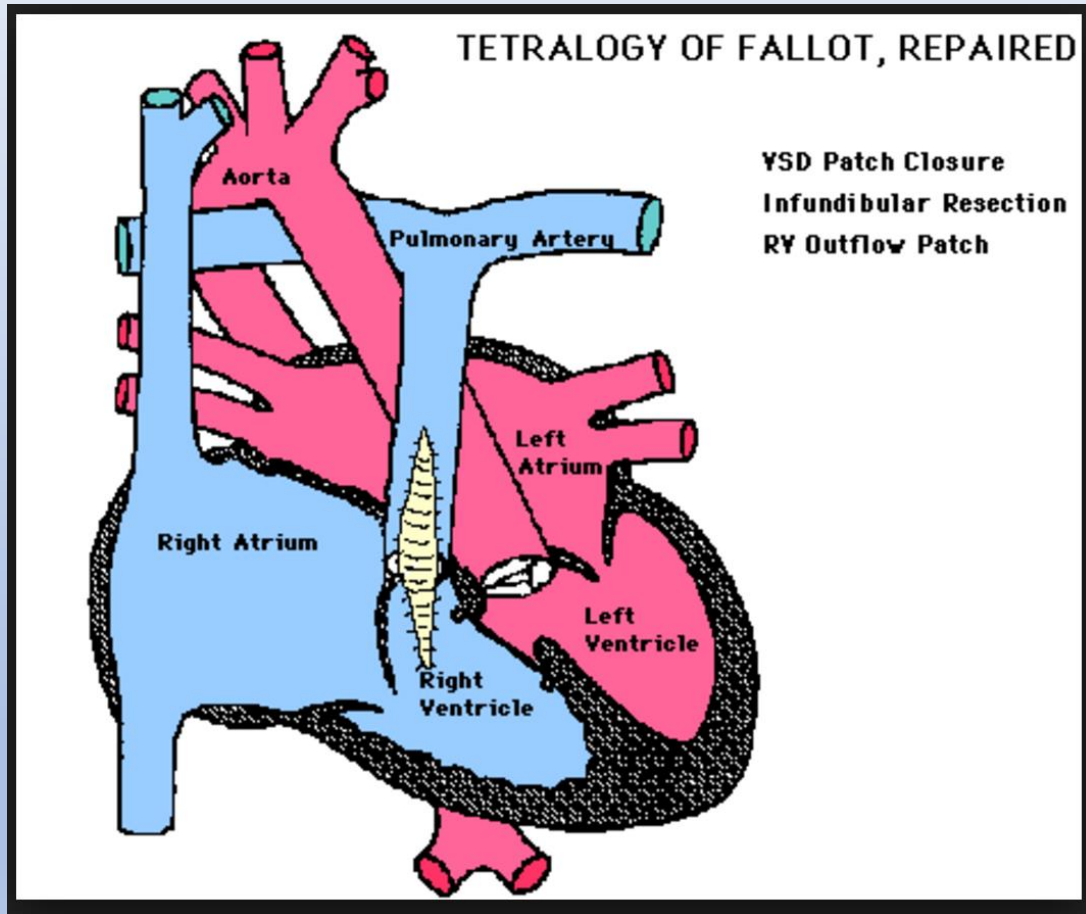
classic

modified



- A landmark operation
- Thoracotomy or sternotomy
- Typically without CPB
- Solves the problem, but
 - Palliative
 - Risk
 - More surgery
- Less and less done as first procedure

Complete repair = fix everything



- Preferred modern treatment
- Excellent results – mortality 1-2%
- One-stop solution
- Key issue = PV

Big enough → keep it ($\approx 60\%$)

Too small → transannular patch (+/- monocusp) or RV-PA conduit (in higher risk cases)

You can live without a PV!

- In high risk cases leave PFO

Controversies

- Low weight spelling baby – palliation or complete repair
- Early routine surgery
- Role of transcatheter palliation – aiming to replace BT shunts in this group, when early correction is too hazardous (cardiac/non-cardiac factors)
 - Balloon dilatation
 - RVOT stenting
- **Questions**
 - Which Tx pathway has lower overall hazard (mortality, morbidity, RV and PA function, freedom from reintervention, long-term results)?
 - Is early correction or transcatheter palliation associated with fewer PV preservations?

Outcomes of surgery → adulthood

- No residual lesions, good function
- Residual problems
 - PS, PR, mixed disease
 - PA stenoses
 - Arrhythmias – atrial, ventricular
 - R-heart hypertrophy and/or dilatation
 - ‘restrictive physiology’
 - Residual VSD, progressive TR or AR
 - If conduit used – too small ± degenerated
 - Dilated Ao root
 - 5 cm ToF ≠ 5 cm Marfan’s

Typical adult patient

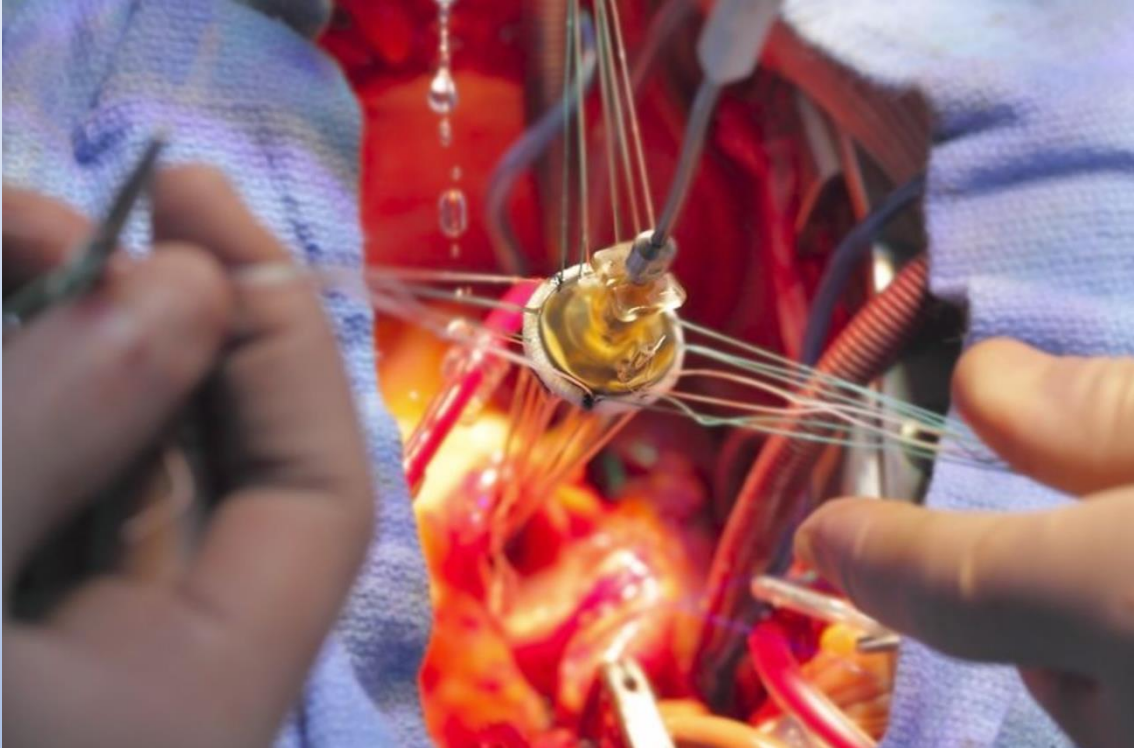
- Young adult
- **Free PR**, dilated RV
- \pm symptoms
 - If asymptomatic do exercise test or operate on RV size criteria
 - Very dilated RV = poor outlook
- Key tests
 - Echo
 - MRI
 - Coronaries if > 40 years – catheter or CT
- Consent process – MDT mandate, shared decision making

Redo PVR / conduit change

- Redo sternotomy
- Careful... everything badly stuck... risk of bleeding ↑



PVR



It is possible to do this on CPB but with the heart beating IF there is no septal communication



Tissue PVR – vastly more common, can be accessed percutaneously in future

Mechanical PVR – reasonable option if patient already on warfarin for another reason (eg MVR)

Conduit change



Hancock valved tube



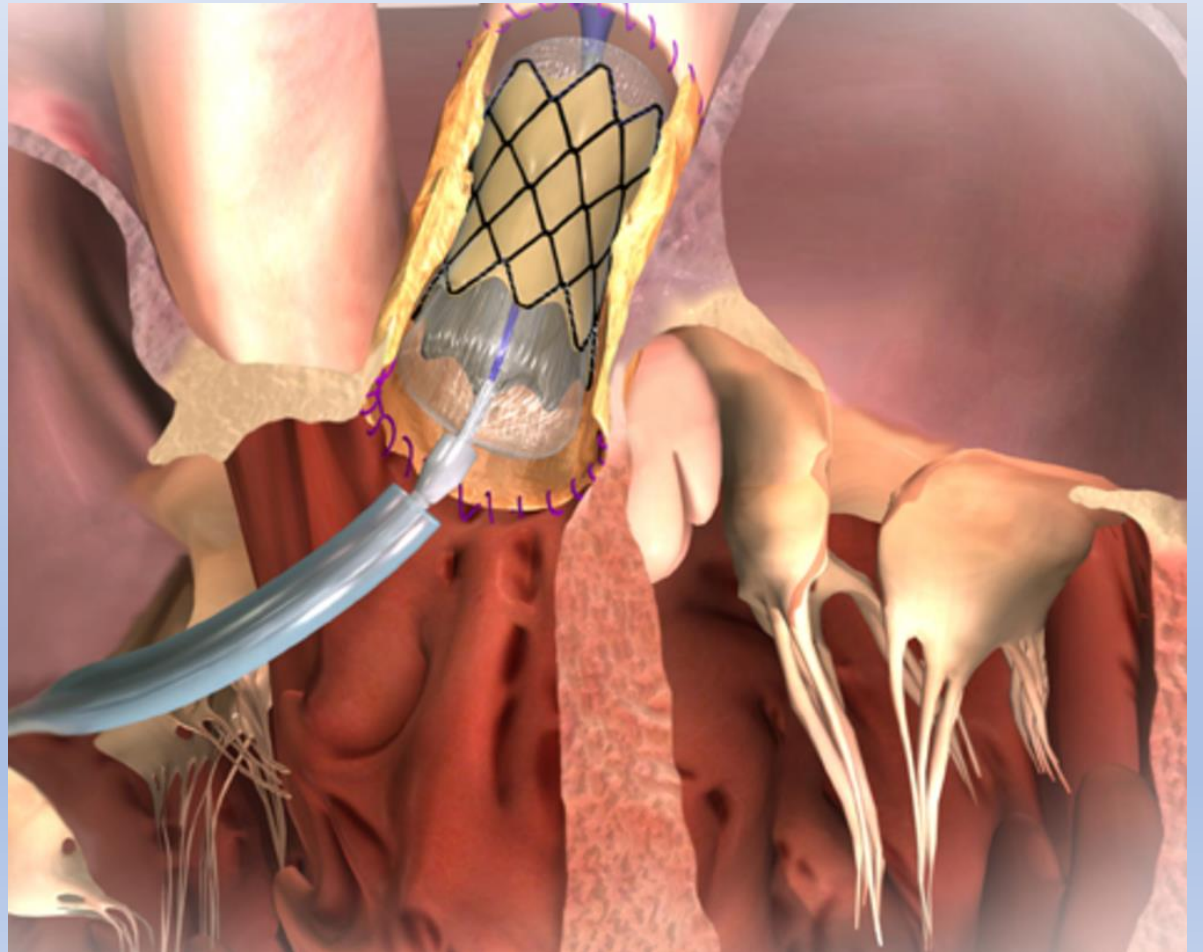
Homograft



Bovine jugular vein (Contegra)

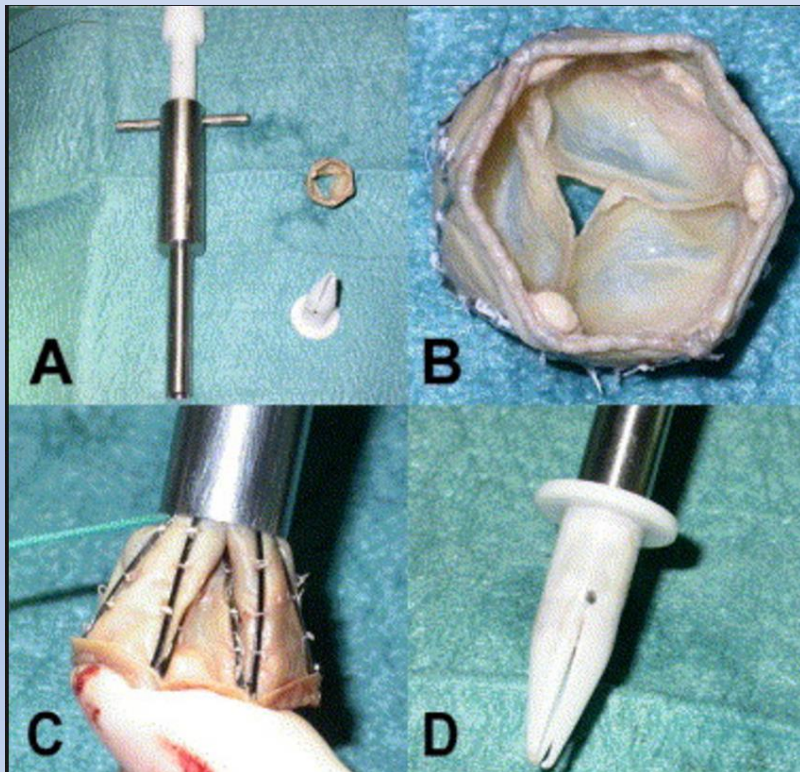
Percutaneous options

- All ages
 - Children – palliation
 - Adults – EP problems, PVR
- Team work – planning, execution, peri-procedure care
- Hybrid



Surgical innovation

- Injectable pulmonary valve, ongoing INVITE study
- Avoids cardiopulmonary bypass
- May need PA plication if too big, not suitable if CPB needed for another reason



Pulmonary valve implantation using self-expanding tissue valve without cardiopulmonary bypass reduces operation time and blood product use

Qiang Chen, MD,^a Mark Turner, MD,^b Massimo Caputo, MD,^a Serban Stoica, MD,^a Stefano Marianeschi, MD,^c and Andrew Parry, DM^a

Other procedures

- PFO/ASD closure
- TV repair / replacement
- Arrhythmia ablation surgery – atria, ventricles
- PA enlargement

- Hybrid work
 - Injectable PV valve – TOE
 - Melody valve – surgical access

Complications, perioperative care

- Mortality – conduit change > PVR, overall 1-3%
- (Bleeding, infection, stroke, MI, pacemaker)

- RV failure – rare, difficult to treat
 - Inotropes
 - Pulmonary vasodilators
 - IABP, ECMO, VAD
- Arrhythmias
- End-organ dysfunction

Age is a saving grace and the majority of patients do well, quick functional recovery

Follow up

... the sensational GUCH liaison service...

- Rehabilitation
- Psychological support
- Anticoagulation, cardiac function
- Return to normal activities

... one of these people has ToF...

