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AD-COR Program inovativ de formare in domeniul cardiologiei pediatrice
POSDRU/179/3.2/S/152012

Data: **14-09-2015**

MODUL TEORETIC

Tetralogy of Fallot

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Activitate prestata de I.R.C.C.S. POLICLINICO SAN DONATO – MILANO, ITALIA in baza contractului nr. 18/22144/29.07.2015

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Acest material a fost documentat/ validat/ prezentat la sesiunile de formare în cadrul proiectului *„AD-COR Program inovativ de formare în domeniul cardiologiei pediatrice”* - POSDRU/179/3.2/S/152012, proiect cofinanțat din Fondul Social Operațional Sectorial Dezvoltarea Resurselor Umane 2007-2013.

Beneficiar: Universitatea de Medicină și Farmacie „Carol Davila” București

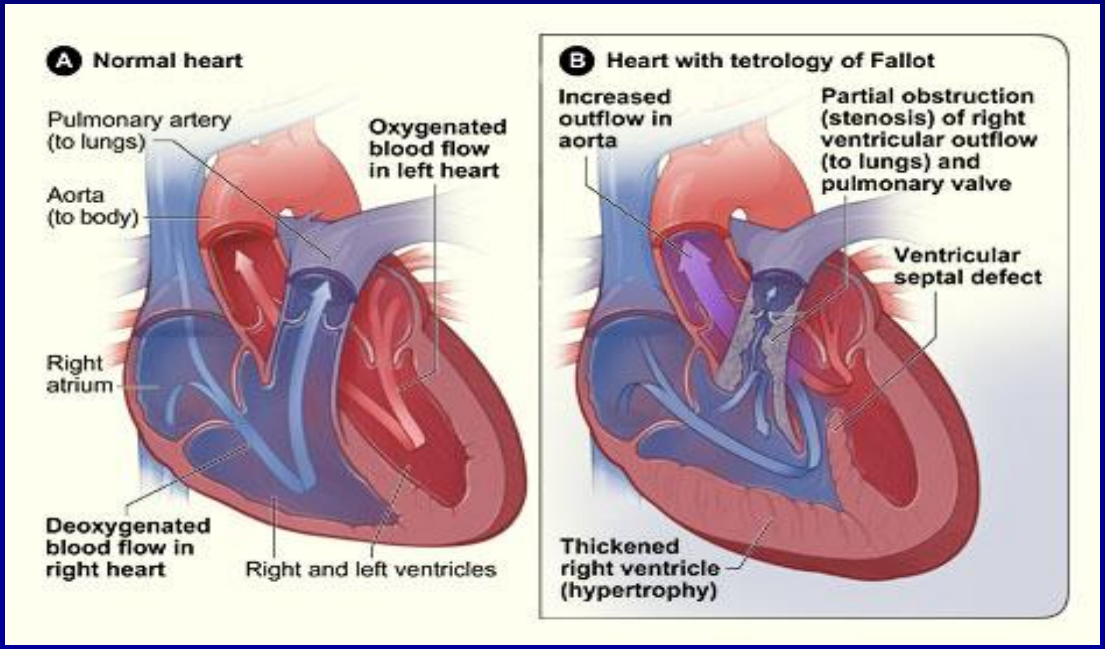
Tetralogy of Fallot

Timing of operation and current techniques

Tammam Youssef

ToF: Definition

- **right ventricular hypertrophy**
- **pulmonary valve stenosis** *(from RVOTO to PA)*
- **transposition of the aorta** *(from minimal to 75%)*
- **ventricular septal defect**



Natural history

ToF occurs in 3 of every 10,000 live births, accounts for 7–10% of all congenital cardiac malformations

Survival without surgery

- 25~35% die in the first year of life
- 40~50% die by the age of 4
- 70% by 10 years
- 95% by 40 years

Natural history

Survival with complete repair

- The outcome after surgical repair of TOF has improved over the last decades and recent studies have reported a mortality rate of 0-7%.

- ＞85% survive to adulthood (> 18 years old)

Expectations from surgery

- minimal early mortality and morbidity
- avoidance of long-term complications and a low probability of early and late reoperations
- good neurodevelopmental and functional status and quality of life
- preservation of pulmonary valve and tricuspid valve function, biventricular contractility

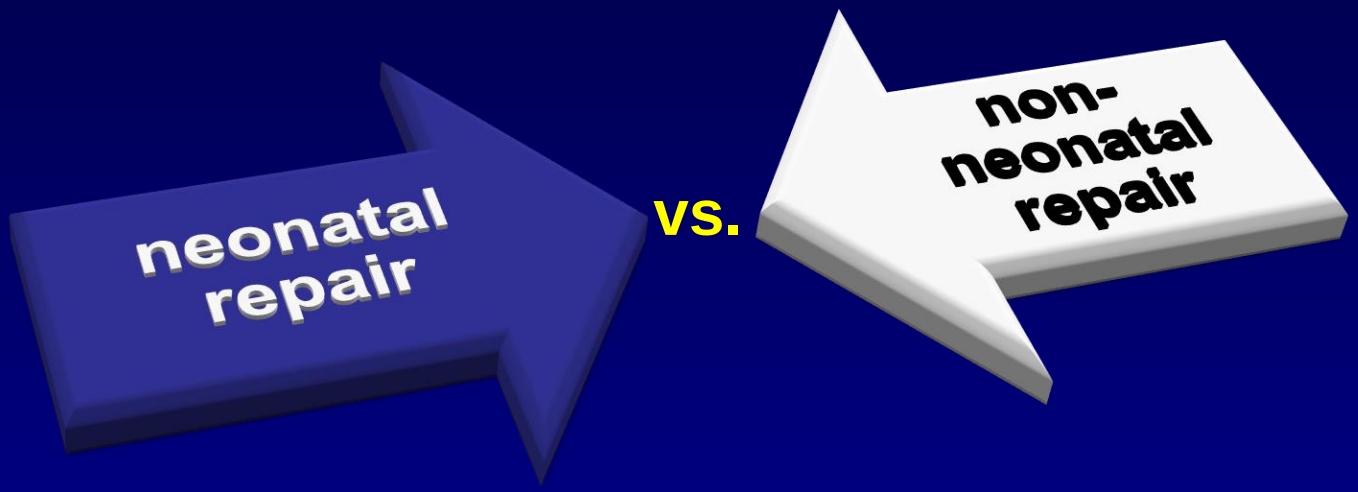
Unsolved issues

(1) the optimal time for repair

(2) early one-stage versus two-stage repair

(3) transatrial or transventricular approach for closure of VSD.

Timing of surgery



The best age for TOF repair remains controversial

*In fact, most tetralogy patients (probably 75%) do not require any surgical treatment in the neonatal period. The important argument therefore is not "primary repair vs shunt" but "**neonatal vs non-neonatal**" elective primary*

Primary neonatal repair

Advantages

- prevention of time related end organ damage from cyanosis
- removal of stimulus for RV hypertrophy and fibrosis
- less wide patch resulting in better right ventricular function and few arrhythmias
- eliminates branch pulmonary artery stenosis or the distortion associated with palliative shunt operation
- improved lung development (vascular and alveolar)
- avoidance of deleterious effects and risks of palliative shunts (shunt thrombosis, CHF,), and psychosocial-economic issues (for the family and care givers).

Primary neonatal repair

Disadvantages

- Newborns with CHD are known to be susceptible to abnormal brain development and widespread CNS abnormalities, which may be exacerbated following neonatal operations utilizing CPB. Studies of neurodevelopmental outcomes of neonates undergoing CPB compared to older children have shown lower intelligence quotients in patients exposed to bypass as neonates (*Miller et al. 1996*).
- Neonatal tetralogy repair strategy often requires an employment of a transventricular approach, with a transannular RVOT patch reconstruction.

Current strategy

- Patients with cyanotic ToF may either undergo neonatal complete repair or neonatal palliation. Peri-operative mortality rates for either surgical approach is less than 3%.
- Surgical management remains dependent on the protocols preferred by the individual centers..
- If neonatal tetralogy surgery can be safely delayed, then non-neonatal primary elective repair (3-6 months) is preferable to a staged procedure.
- In symptomatic young infant less than 3 months old, a palliative shunt would be considered and performed initially to avoid the morbidity of prolonged ventilation and hospital stay.

Surgical Techniques

Surgical Approach

- **Palliative procedures**
- **Complete repair**
  - Goals
    - Maximal relief of RVOTO
    - Closure of VSD
    - Preservation of RV function
  - Full correction between age of 2 to 10 months
    - Primary total repair
    - Staged surgery
      - Presence of coronary abnormalities
      - Multiple VSDs
      - Inadequate pulmonary artery anatomy

Induction agents

- Intravenous
  - Ketamine: maintaining SVR
  - Barbiturate: ↓both SVR and PVR
- Inhalation
  - Sevoflurane: least effect on SVR } both safe
  - Halothane }
  - Desflurane } dose-dependent ↓SVR
  - Isoflurane } pungent odor
  - Nitrous oxide

Operative technique

- CPB is established (28-31°C) through bicaval and aortic cannulation. Systemic to PA shunts, if present, are ligated after the beginning of CPB. The heart is arrested with antegrade cold blood cardioplegia. The VSD is closed through the RA or RV. The foramen ovale is closed, the heart is de-aired, and the clamp is removed.

- We utilize either **transatrial** or **transventricular** approach.

Transatrial approach

- RA is opened longitudinally, and the left heart is vented through the foramen ovale or ASD. Excision of the parietal extension of the infundibular septum is performed through the TV and RV as required. Hegar dilators are passed either through the TV or RV toward the main PA. If the annular Z score is < –2 (referenced to predicted norm for the patient's weight) the annulus is divided.

- **Advantages:**

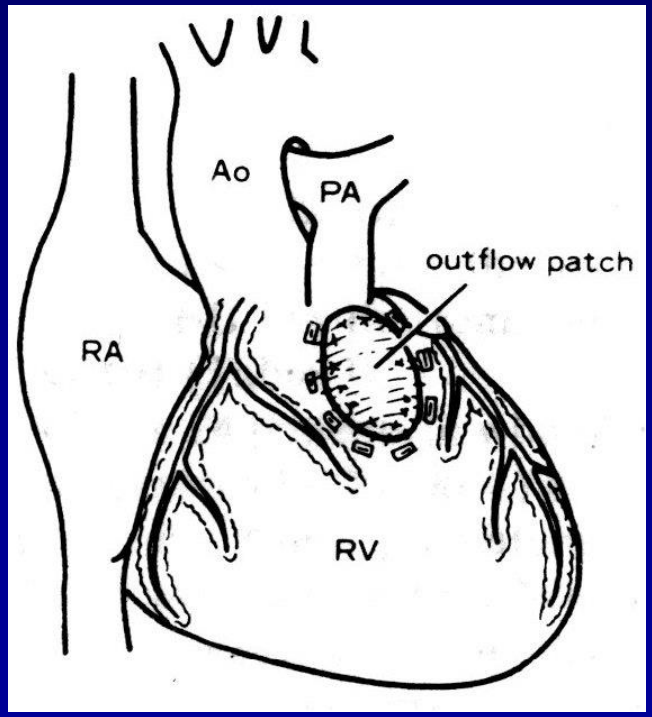
- The long-term function of the RV is preserved
- The risk of injuring coronary artery is minimized.
- The resultant pulmonary regurgitation after limited transannular patching is less severe than that which often occurs after transventricular repair.

Transventricular approach

RV incision for access to both the RVOTO and ventricular septal defect (VSD). The relatively favorable long term results have been noted in numerous studies.

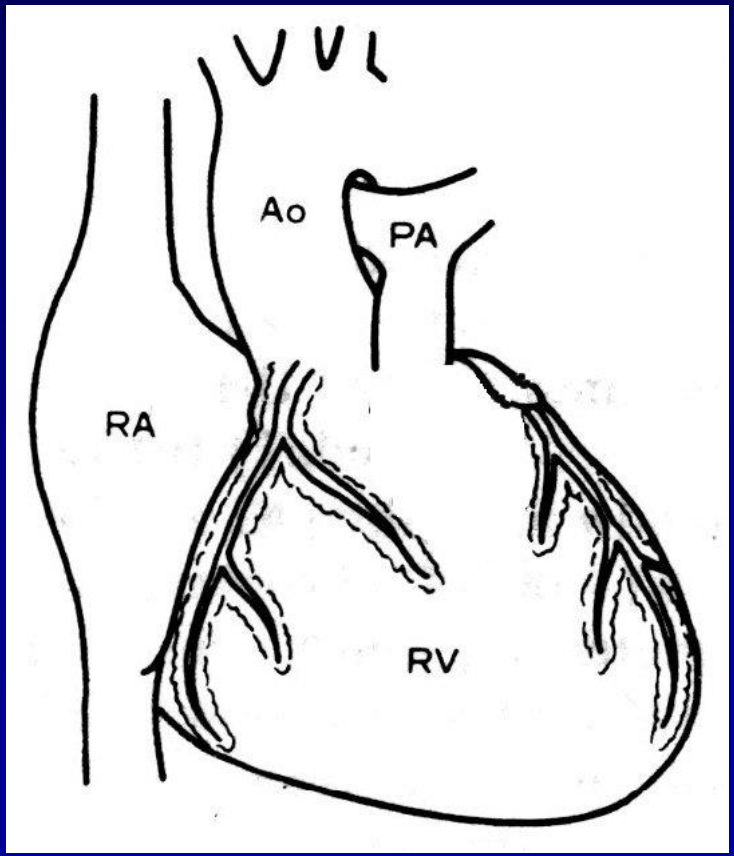
❖ **Disadvantages:**

- myocardial injury
- coronary injury
- RV dysfunction
- arrhythmias related to the right ventriculotomy



Transpulmonar approach

A longitudinal incision is made in the main PA and extended to the annulus of the PV.

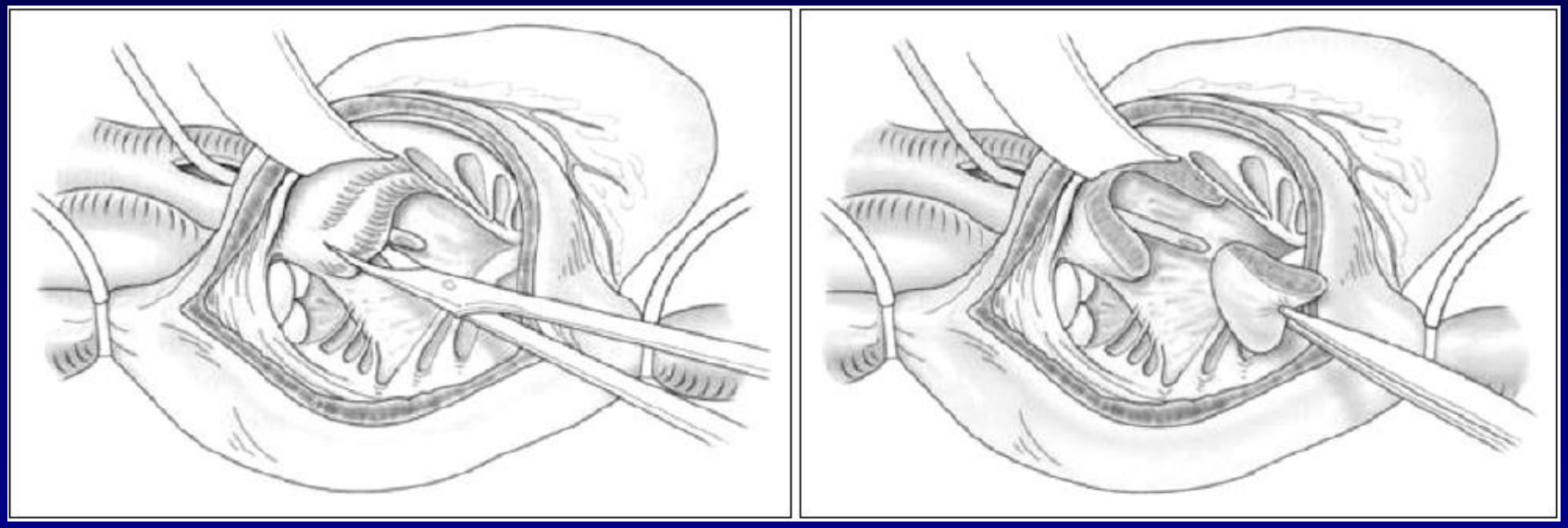


TA-TP approach

The TA-TP approach involves RV outflow tract (RVOT) resection via the TV and PV, and VSD closure via TV. If need a limited ventriculotomy (outlet portion of RV or infundibulum), which is reconstructed with a transannular patch. The TA-TP approach has resulted in significantly less RV dilatation and better preservation of contractility at 10 years follow up.

Karl et al. concluded from their results that the TA-TP approach resulted in a similar operative risk compared with transventricular TOF repair, with a 97.5% actuarial survival rate at 42 months.

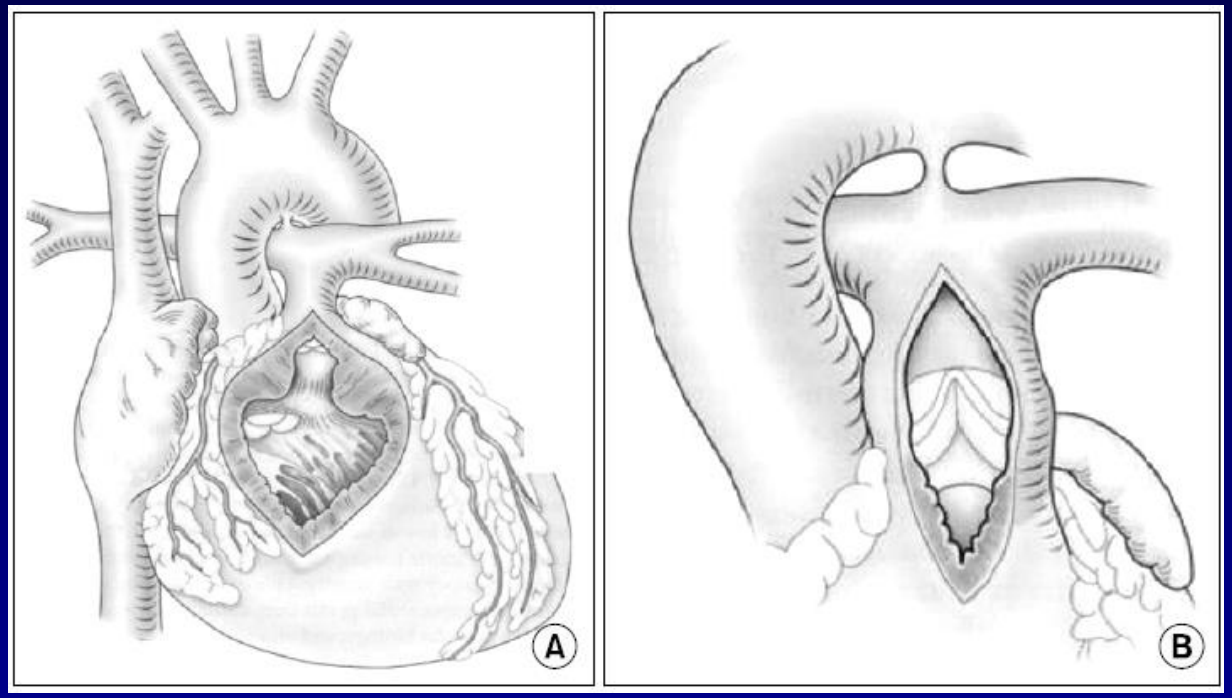
TA-TP approach



Resection of the parietal extension of the infundibular septum, using a transtricuspid approach. A Hegar dilator has been passed from the pulmonary artery into the right ventricular (RV) to demonstrate the location of the RV outflow tract

(Karl TR. Ann Pediatr Card 2008)

TV and TA-TP approach



Comparison of the classical **transventricular** approach (A) and the **transatrial-transpulmonary** approach (B). In the latter strategy the right ventricular (RV) incision is limited to what is required to relieve the RVOTO, with VSD closure and RVOT resection performed via the tricuspid valve and pulmonary valve

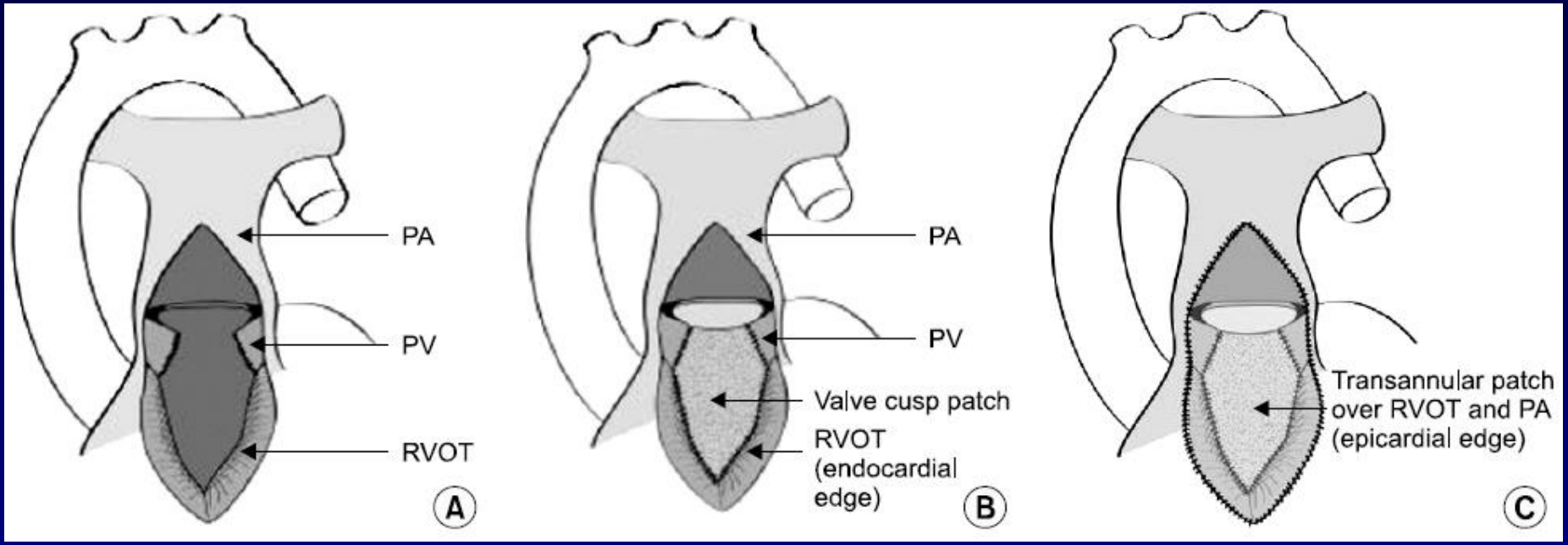
(Karl TR. Ann Pediatr Card 2008;1:93-100)

Surgical complications

- pulmonary regurgitation
- recurrent or residual pulmonary stenosis
- ventricular arrhythmias

Strategies to limit the size of the right ventriculotomy and reduce the incidence of PI:

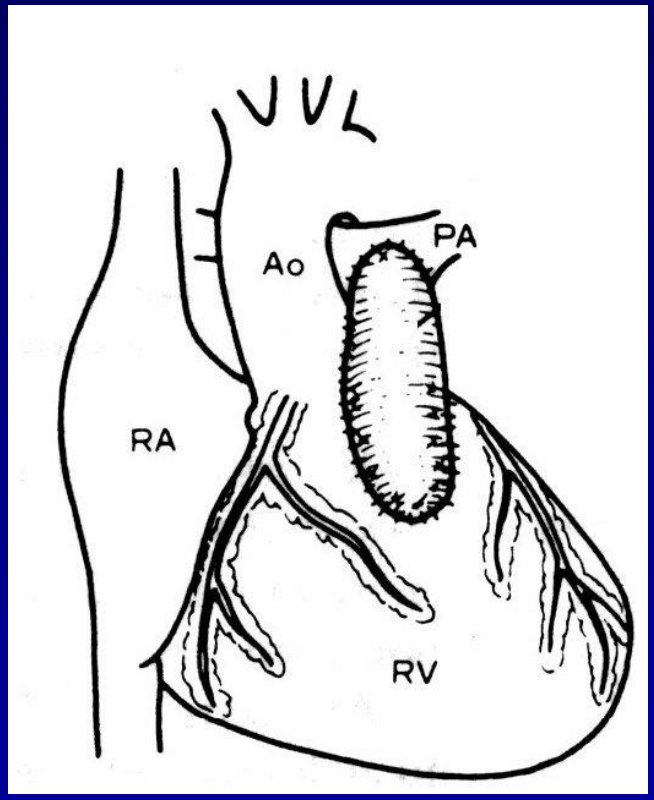
- Valved homografts and xenografts (**BUT** eventually need replacement).
- Monocusp valve created from pericardium, xenograft valve cusps, fascia lata, autologous pulmonary artery wall, or PTFE (decrease short-term PI, **BUT** have limited durability).



Pulmonary cusp augmentation repair technique.
(A) A transannular incision has been created according to calibration of the right ventricular outflow tract (RVOT) diameter. The incision divides the anterior pulmonary valve leaflet, although the exact location will vary with the valve orientation and morphology. (B) A triangular patch of glutaraldehyde-preserved autologous pericardium is sutured to the epicardial edge of the RVOT incision and to the divided edges of the valve leaflet. The leaflet dimensions are based on the caliber of a normal pulmonary valve diameter and should provide sufficient free edge diameter to ensure coaptation with the native valve remnant. (C) A second larger oval patch of the same material is sutured to the epicardial aspect of the RVOT incision and to the edges of the pulmonary arteriotomy, creating a sinus anterior to the reconstructed leaflet. PA, pulmonary artery; PV, pulmonary valve. (Karl TR. Ann Pediatr Card 2008;1:93-100)

Transanular patch

Transannular patches (across the ventriculopulmonary junction), create a state of chronic pulmonary regurgitation, which increases morbidity in young adults, producing exercise intolerance and ventricular arrhythmias.



Transanular patch

- The incidence of transannular patch use at the time of complete repair **in younger patients ranges from 90 to 100%** (***Touati et al. Primary repair of tetralogy of Fallot in infancy. J Thorac Cardiovasc Surg. 1990.***
  Pozzi et al.Tetralogy of Fallot: what operation, at which age. Eur J Cardiothorac Surg. 2000).
- A report reviewing 56 patients with TOF less than 6 months of age showed a significantly lower incidence of transannular patching in the patients who were initially palliated with a shunt (9% transannular patches) compared with 41 patients who underwent initial complete repair (**56% transannular patches**). (**Sousa et al. Surgery for tetralogy of Fallot at less than six months of age. J Thorac Cardiovasc Surg 1994**)

Transanular patch - Complications

- Several studies showed that correction in infancy carries a higher risk of TAP (*Knott-Craig et al. A 26 year experience with surgical management of Tetralogy of Fallot: risk analysis for mortality or late reintervention. Ann Thorac Surg 1998*).

- Some previous studies have also shown that age at operation less than about 6 months, low weight, and **TAP reconstruction** of the RVOT are all associated with increased mortality after intracardiac repair.

Transanular patch - Complications

Harald L et al. demonstrated that a significant worse outcome is registered in the TAP group (*Harald et al. Single-center 50 years' experience with surgical management of tetralogy of Fallot. EJCTS 2011*).

d'Udekem et al. also reported that TAP and RV patching equally affect late functional status (*d'Udekem et al. Tetralogy of Fallot: transannular and right ventricular patching equally affect late functional status. Circulation 2000).*

Outcome after repair of tetralogy of Fallot in the first year of life (literature review)

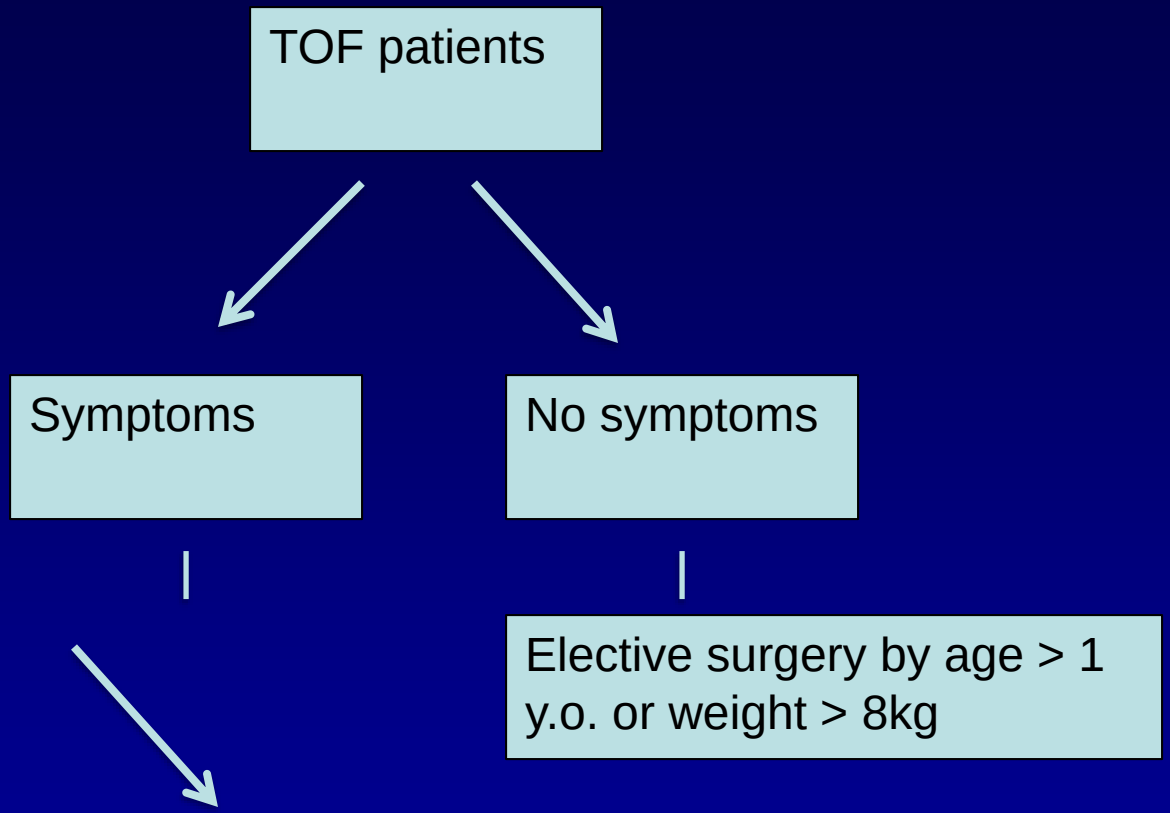
Author	Year of Publication	No of patients	Median Age (months)	Hospital mortality N (%)
Hennein H.A.	1995	30	0,4	0
Kaushal S.K.	1996	52	10,2	3 (5,6%)
Underleider M.	1997	22	3,4	0
Knott-Craig C.	1998	63	7,2	3 (4,8%)
Caspi J.	1999	82	5,2	0
Pigula F.A.	1999	99	1,1	3 (3%)
Reddy V.M.	1999	120	1,9	2 (1,6%)
Rao V.	2000	50	9	0
Alexiou C.	2001	89	6,3	1 (1,1%)
Lazorychynets VV	2001	74	9,9	5 (6,8%)
Cobanoglu A.	2002	63	6,5	4 (6,3%)
Rachmat J,	*2004*	*40*	*10,4*	*2 (5%)*

Conclusions

- Patients undergoing primary repair at 3 months of age. Have length of intensive care unit stay, period of mechanical ventilation and the need for inotropes is increased

- Closing the VSD through the infindibulotomy is feasible and safe. It avoids right atriotomy and potential risk of injury of the tricuspid valve.

- The upmost pathology of ToF is the obstruction of the RVOT (infundibulum). Therefore, there **can not be** repair of ToF staying away from infindibulum.


TOF patients
Symptoms
No symptoms
Elective surgery by age > 1 y.o. or weight > 8kg