Past current and optimal surgical management of tetralogy of Fallot

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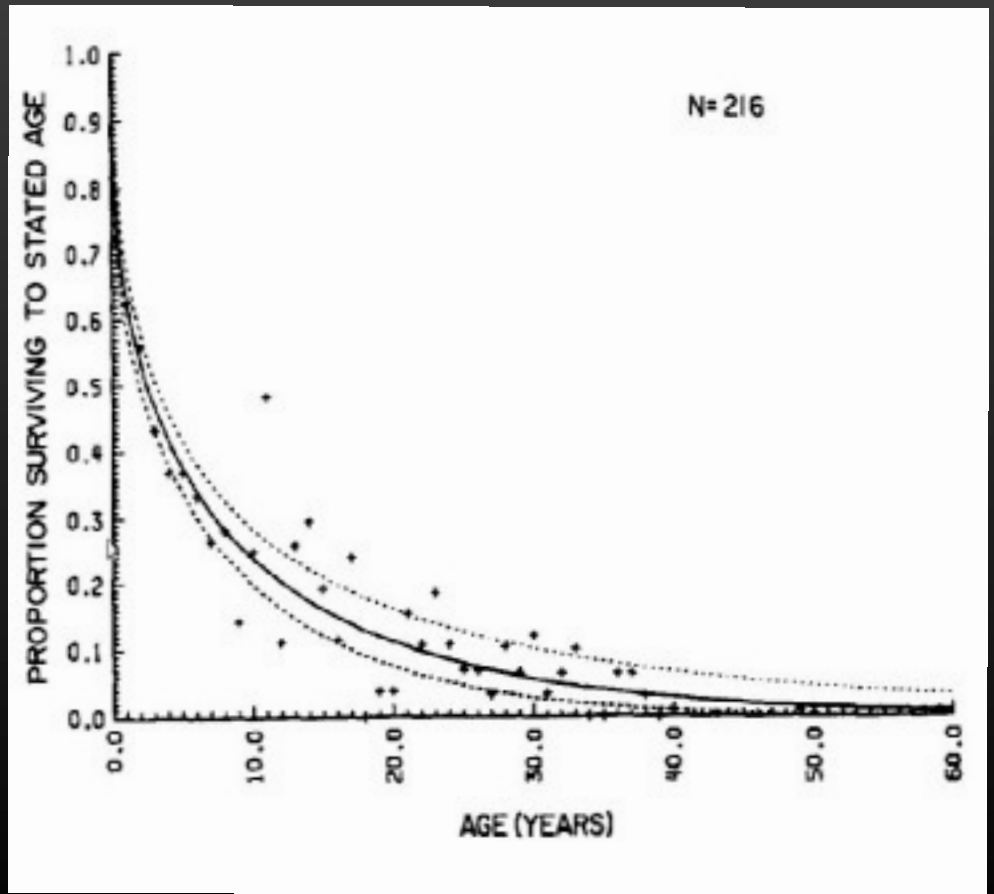
NHS Foundation Trust

Why do we repair ToF?

Life expectancy of persons with ToF
based on data from the Danish population study



Hypoxic spells
Cerebral accidents and abcesses

Bertranou et al, AJC 1978
Rygg et al, Dan Med Bull 1971

Repair of Tetralogy of Fallot The beginning (1944)

- 1944: Blalock Taussig Shunt
  - in 1941 Taussig said to Blalock, "I stand in awe and admiration of your surgical skill—but a truly great day will come when you can build a ductus for a cyanotic child, not when you close a ductus for a child who has a little too much blood going to his lungs." Blalock replied, "When that day comes, this will be child's play" (Taussig HB (1979) Neuhauser lecture—tetralogy of Fallot: early history and late results. AJR Am J Roentgenol 133:423–431)

A little over 50% of the original patients who survived the initial operation were still alive at 20 years follow-up

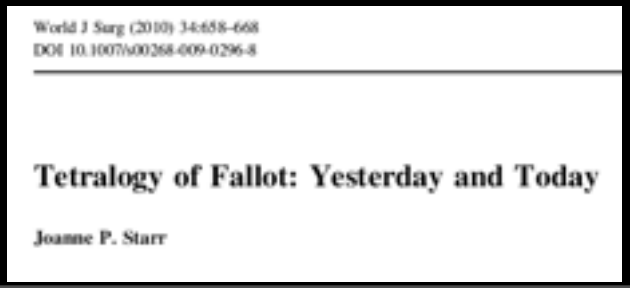
Repair of Tetralogy of Fallot Early years (1950s)

Mortality in the early years of repair was as high as 60%, but by the early 1960s perioperative mortality decreased significantly to 7%–14%. Ages of these patients ranged from 15 months to 54 years. The mortality rate was higher in patients with severe cyanosis and younger age. **The number of patients who had previous shunts ranged from 43% to 75%**

Kirklin JW, Payne S, Theye RA et al (1960) Factors affecting survival after open operation for tetralogy of Fallot. Ann Surg 152:485–491

Kirklin JW, Wallace RB, McGoon DC et al (1965) Early and late results after intracardiac repair of tetralogy of Fallot. Ann Surg 162:578–587

Azar H, Hardesty RL, Pontius RG et al (1969) A review of total correction in 200 cases of teralogy of Fallot. Arch Surg 99: 281–285



Wolf MD, Landtman B, Neill CA et al (1965) Total correction of tetralogy of Fallot I. Follow-up study of 104 cases. Circulation 31:385–393

Goldman BS, Mustard WT, Trusler GS (1968) Total correction of tetralogy of Fallot—review of ten years' experience. Br Heart J 30:563–568

Ebert PA, Sabiston DC (1967) Surgical management of the tetralogy of Fallot: influence of a previous systemic-pulmonary anastomosis on the results of open correction. Ann Surg 165:806–812

Gerbode F, Johnston JB, Sader A et al (1961) Complete correction of tetralogy of Fallot. Arch Surg 82:793–800

Repair of Tetralogy of Fallot

First series reported in the late 1960s

Recommendations:

Repair considered after 5 years of age

Shunt below 5 years with elective repair between 8-12

Survival 85-90% by the end of the 1970s

Barratt-Boyes BG, Neutze JM (1973) Primary repair of tetralogy of Fallot in infancy using profound hypothermia with circulatory arrest and limited cardiopulmonary bypass: a comparison withconventional two stage management. Ann Surg 178:406–411

Pacifico AD, Bargeron LM Jr, Kirklin JW (1973) Primary total correction of tetralogy of Fallot in children less than four years of age. Circulation 48:1085–1091

Repair of Tetralogy of Fallot

First series late 1960s

Technique

- Repair of the ventricular septal defect via a large right ventriculotomy
- Extensive resection of the right ventricular outflow musculature and pulmonary valve leaflets.

Long term

- Pulmonary regurgitation
- Dilatation of the RV
- Aortic insufficiency

Repair of Tetralogy of Fallot Recent era from 1980s

Earlier repair as young as within the first 3 months :

- Avoid exposure of pulmonary vasculature to low pressure and low flow and RV Hypertrophy with decrease of compliance and arrhythmias
- Avoid chronic cyanosis
- Economic and psychosocial advantage

Mortality as low as 4%

DiDonato RM, Jonas RA, Lang P et al (1991) Neonatal repair of tetralogy of Fallot with and without pulmonary atresia. J Thorac Cardiovasc Surg 101:126–137

Repair of Tetralogy of Fallot
Concerns regarding long term results

Right ventricle

- Dilatation, Function
- Consequences of pulmonary valve insufficiency?

Arrythmias and sudden death

- Late repair? Ventriculotomy? hypertrophy, fibrosis or Dilatation of the Right ventricle and right atrium?

Aortic root dilatation

- Previous shunts
- Late repair

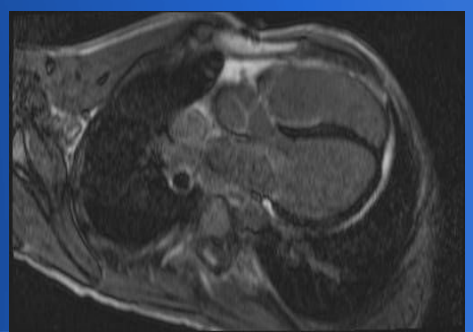
Reinterventions

- Linked with use of a transannular patch?

Pulmonary regurgitation after repair of ToF: Not a benign condition

- Volume overload of the RV
- RV dilation
- RV dysfunction
- RV fibrosis
- RV restriction

- RV-LV interaction
- LV dysfunction
- Exercise intolerance
- QRSd prolongs
- Arrhythmias, SCD



TOF patients born ~30 years ago adult ToF patients today

- Low perioperative mortality
- High prevalence of pulmonary regurgitation
- Arrhythmias
- Sudden cardiac death
- Exercise intolerance - Heart failure
- Other: Pregnancy, endocarditis, aortopathy, RVOTO, TR

Optimisation of Repair of Tetralogy of Fallot

Preservation of the Right ventricle

Kawashima Y, Kitamura S, Nakano S et al:Corrective surgery for ToF without or with minimal right ventriculotomy and with repair of the pulmonary valve. Circulation 64/II.147-II153, 1981

Transatrial-Transpulmonary Repair of Tetralogy of Fallot

Massimo A. Padalino, Vladimiro L. Vida, and Giovanni Stellin

Pediatric Cardiac Surgery Annual

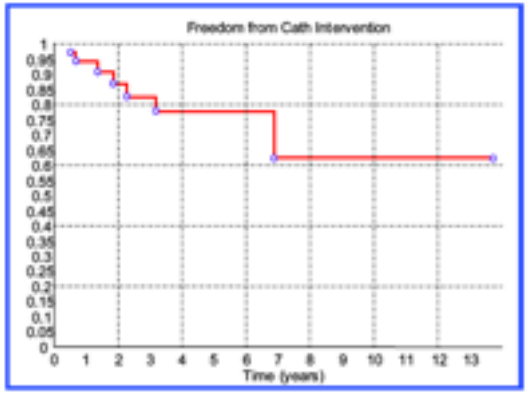


Figure 8 Freedom from reoperation in the subgroup of patients operated within 3 months of life.

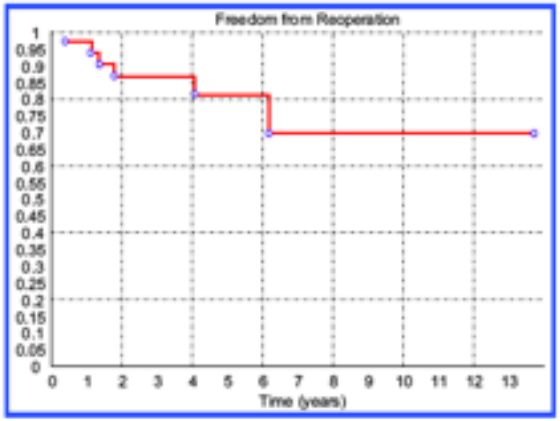


Figure 7 Freedom from reoperation in the subgroup of patients operated within 3 months of life.

Optimisation of Repair of Tetralogy of Fallot
Avoidance of neonatal palliation?

- Reasons:
  - Morbidity and mortality related to palliation
    - Shunt mortality 7% in the Toronto series
  - Longer exposure to hypoxia, cyanosis and high pressures

Chowdhury UK, Sathia S, Ray R, Singh R, Pradeep KK, Venugopal P. Histopathology of the right ventricular outflow tract and its relationship to clinical outcomes and arrhythmias in patients with tetralogy of Fallot. *J Thorac Cardiovasc Surg* 2006; 132: 270–77.

  - Palliation= another operation?

New palliation options

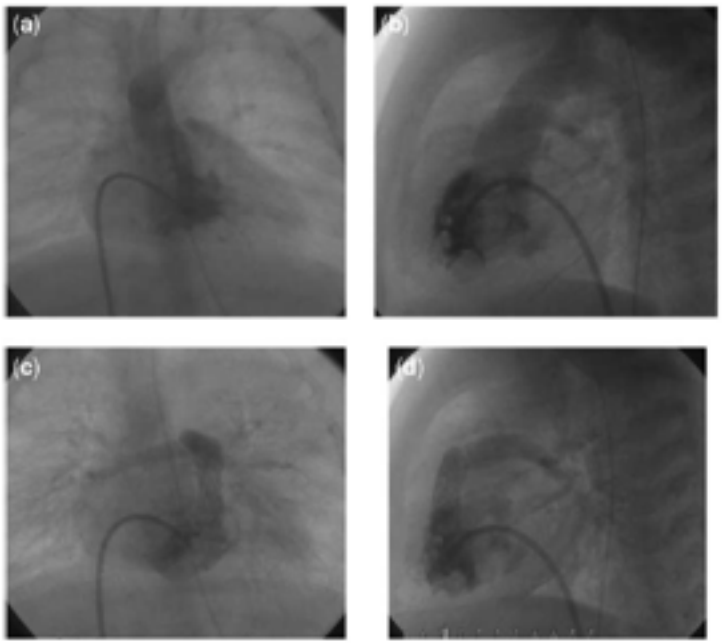


Figure 2.
(a) Patient 2. Right ventricular angiography, frontal view. There is predominant blood flow to the aorta via the VSD and only diminutive flow to the pulmonary arteries. (b) Patient 2. Right ventricular angiography, lateral view. There is predominant blood flow to the aorta via the VSD; the RVOT is completely obstructal and there is only diminutive flow to the pulmonary arteries. (c) Patient 2. Right ventricular angiography, frontal view. After RVOT stening (Palmaz® Blue™), blood flow is now directed to the pulmonary arteries. (d) Patient 2. Right ventricular angiography, lateral view after stent implantation (Palmaz® Blue™). There is adequate antegrade perfusion across the right ventricular outflow tract. The pulmonary valve is distal to the stent. RVOT = right ventricular outflow tract; VSD = ventricular septal defect.

Cardiology in the Young (2013); Page 1 of 5

Repair of Tetralogy of Fallot
The problems with neonatal repair

- Higher morbidity below 3 months of age
- Neurological sequellae
- More and larger transannular patches?

Ooi A, Moorjani N, Baliulis G, et al: Medium term outcome for infant repair in Tetralogy of Fallot: Indicators for timing of surgery. Eur J Cardiothorac Surg 30:917-922, 2006

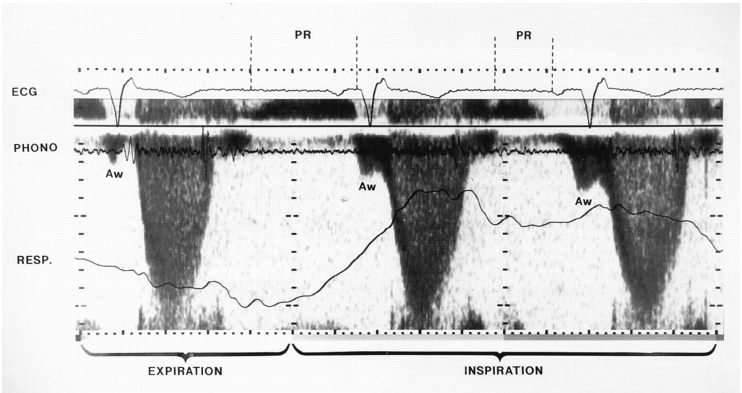
Van Arsdell GS, Maharaj GS, Tom J, et al: What is the optimal age for repair of tetralogy of Fallot? Circulation 102:III123-III129, 2000

Van Dongen EI, Glansdorp AG, Mildner RJ, et al: The influence of perioperative factors on outcomes in children aged less than 18 months after repair of tetralogy of Fallot. J Thorac Cardiovasc Surg 126:703-710, 2003

Repair of Tetralogy of Fallot
The transannular patch and pulmonary valve preservation issue

Gatzoulis MA, Clark AL, Cullen S, Newman CG, Redington AN. Right ventricular diastolic function 15 to 35 years after repair of tetralogy of Fallot. Restrictive physiology predicts superior exercise performance. *Circulation* 1995; 91: 1775–81.

Doppler examination of pulmonary arterial flow in a patient with restrictive right ventricular physiology.



Gatzoulis M A et al. Circulation 1995;91:1775-1781

American Heart Association
Learn and Live

Repair of Tetralogy of Fallot
The transannular patch and pulmonary valve preservation issue

Long-term results of right ventricular outflow tract reconstruction in neonatal cardiac surgery: Options and outcomes

Aditya K. Kaza, MD, Hong-Gook Lim, MD, Daniel J. Dibardino, MD, Victor Bautista-Hernandez, MD, Joshua Robinson, MD, Catherine Allan, MD, Peter Laussen, MBBS, Francis Fynn-Thompson, MD, Emile Bacha, MD, Pedro J. del Nido, MD, John E. Mayer, Jr, MD, and Frank A. Pigula, MD

The Journal of Thoracic and Cardiovascular Surgery • Volume 138, Number 4 911

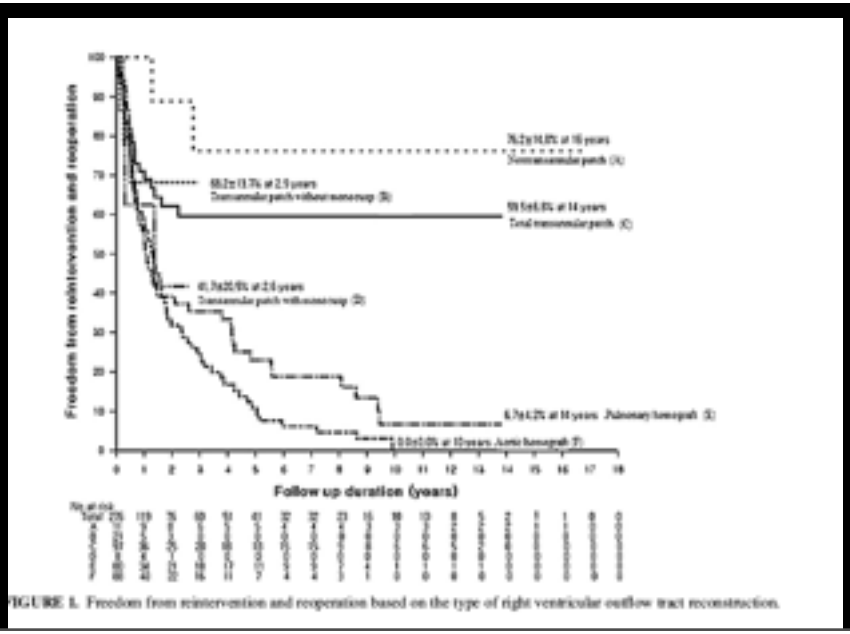
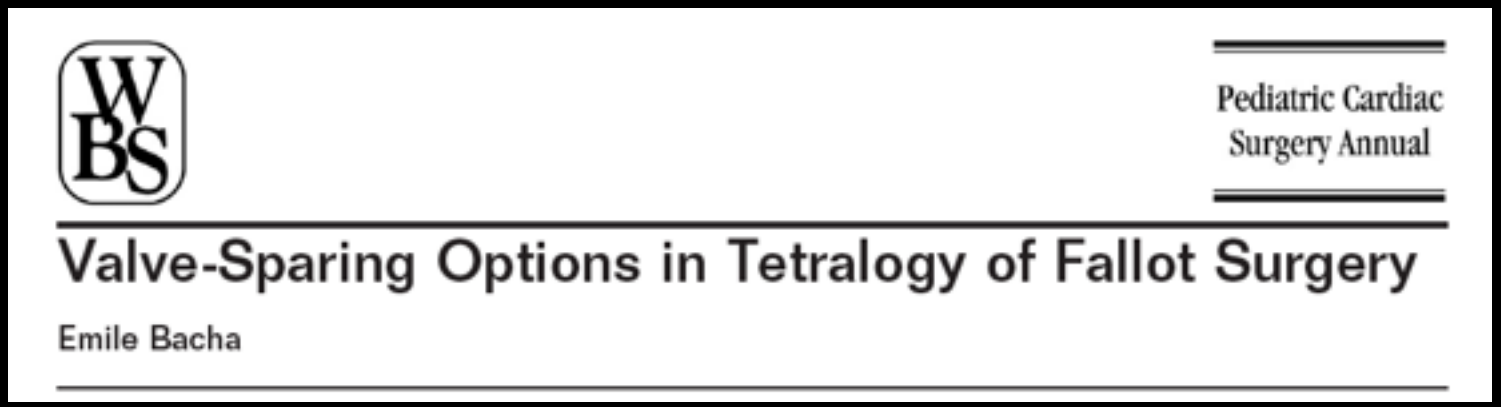
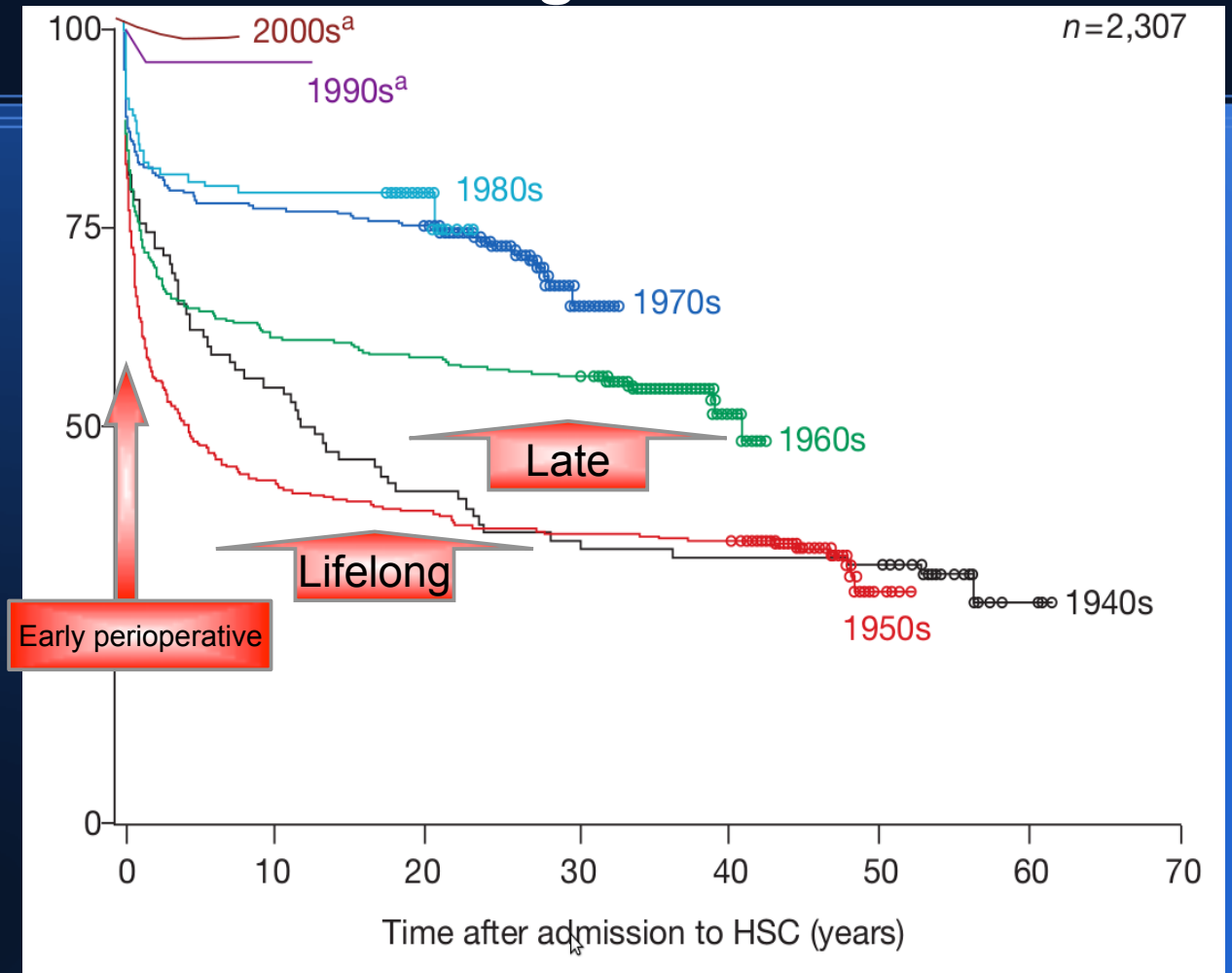


FIGURE 1. Freedom from reintervention and reoperation based on the type of right ventricular outflow tract reconstruction.

Repair of Tetralogy of Fallot
The transannular patch and pulmonary valve preservation issue



3 phases of risk in TOF according to time of diagnosis



Karamlou et al. Nature clinical practice. November 2006

Repair of Tetralogy of Fallot
Today's paradigm

No early mortality, low post-operative morbidity

No life long attrition rate

No long term complications

Preserve the right Ventricle

Strategy?

Repair of Tetralogy of Fallot
Today's paradigm

Tetralogy of Fallot Repair: The Right Ventricle Infundibulum Sparing (RVIS) Strategy

David L. Morales, Farhan Zafar, and Charles D. Fraser, Jr

Because we are not focused on age, what is the optimal timing for a RVIS TOF repair? We feel that optimal timing for complete TOF repair is when one can minimize: (1) the right ventriculotomy; (2) neurological sequelae; (3) need for re-operations; (4) peri-operative morbidity and mortality; and (5) when one can preserve the native pulmonary valve.

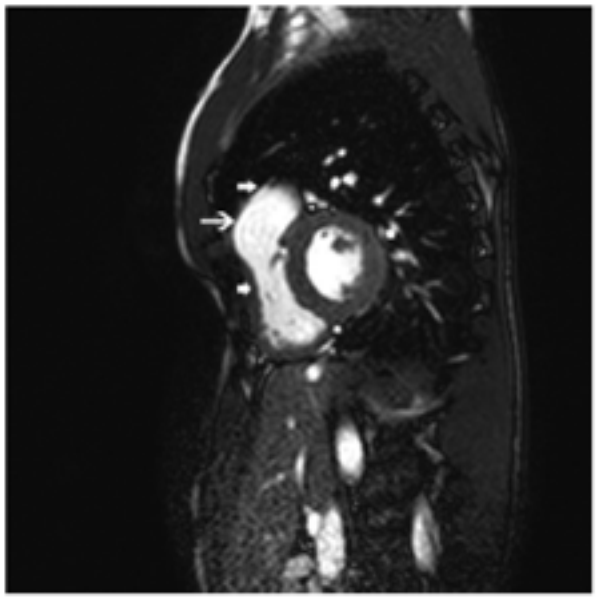


Figure 1 Akinetic infundibulum. *Bold arrows* point to the extent of infundibular patch; *Long arrow* points to the pulmonary valve location.

Tetralogy of Fallot Repair: The Right Ventricle Infundibulum Sparing (RVIS) Strategy

David L. Morales, Farhan Zafar, and Charles D. Fraser, Jr

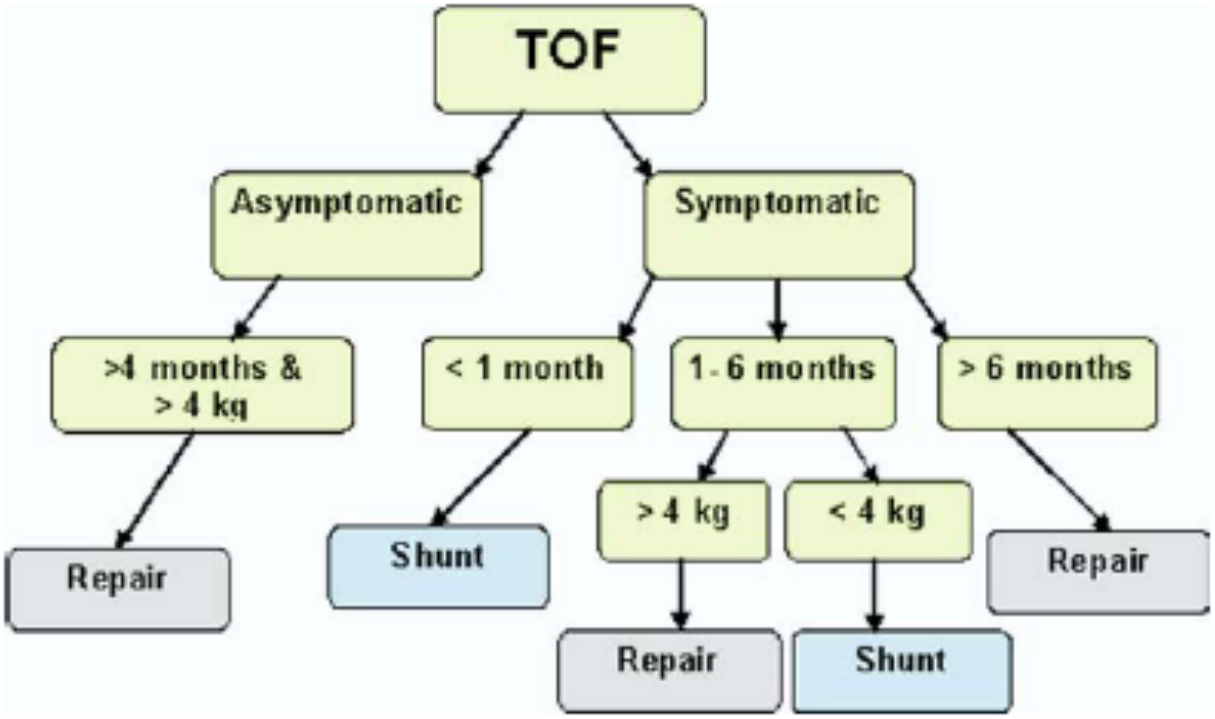


Figure 3 Flow diagram to illustrate the strategy of TOF repair.

Repair of Tetralogy of Fallot Patient tailored approach and clinical pathways

The optimal approach should be tailored to the patient and to the centre capabilities

An integrated care pathway determines locally agreed multidisciplinary practice, based on guidelines and evidence where available for a specific patient/client group...

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Fetal diagnosis of Tetralogy of Fallot

Management of the newborn with CHD starts with ACCURATE prenatal diagnosis based on which APPROPRIATE & OPTIMAL management can be achieved

Julene S Carvalho

Consultant Fetal and Paediatric Cardiologist

Reader in Fetal Cardiology

Tetralogy of Fallot

Associated abnormalities - common

- Chromosomal abnormalities
  - 22q11 microdeletion
  - trisomy 21

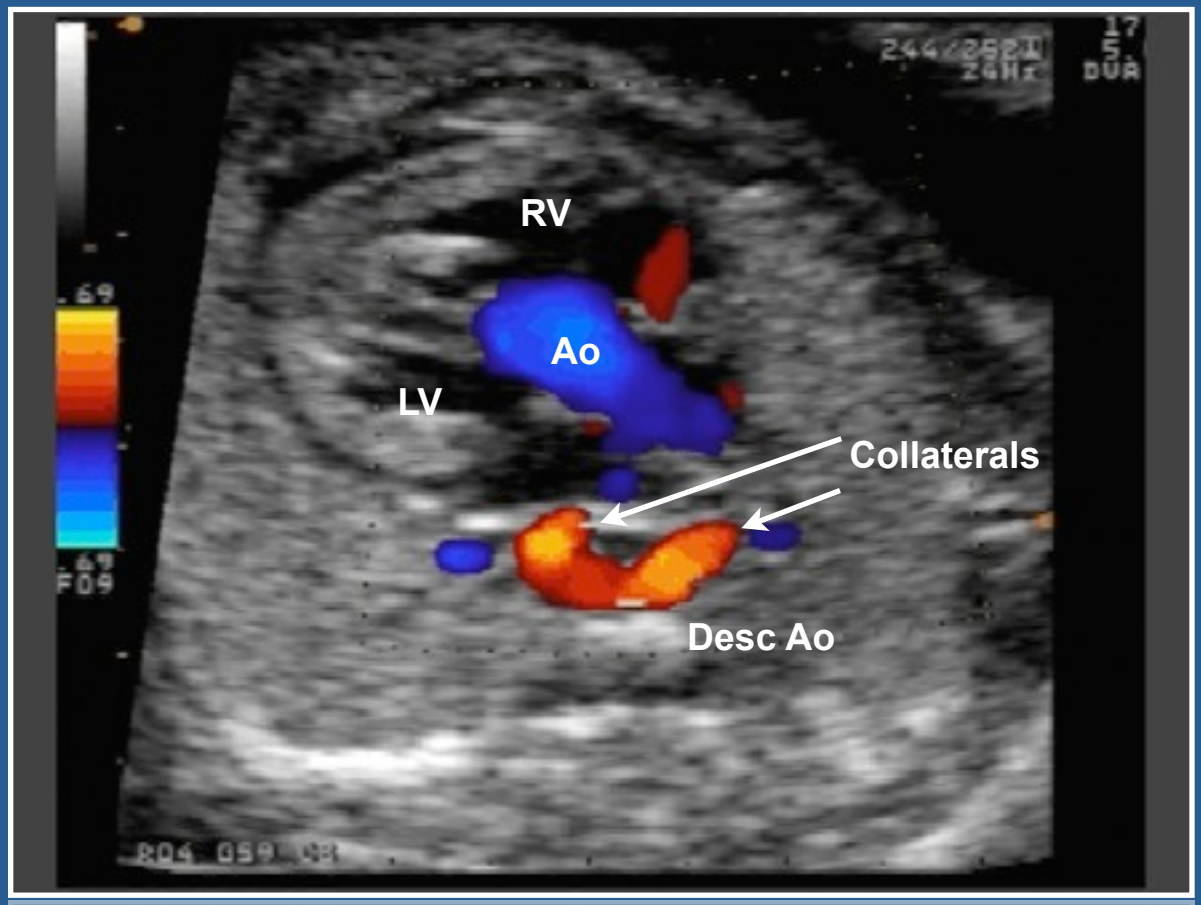
Tetralogy of Fallot

Associated abnormalities - common

- Chromosomal abnormalities
  - 22q11 microdeletion
  - trisomy 21

- Extracardiac abnormalities
  - GI obstruction
  - Exomphalos
  - Diapragmatic hernia

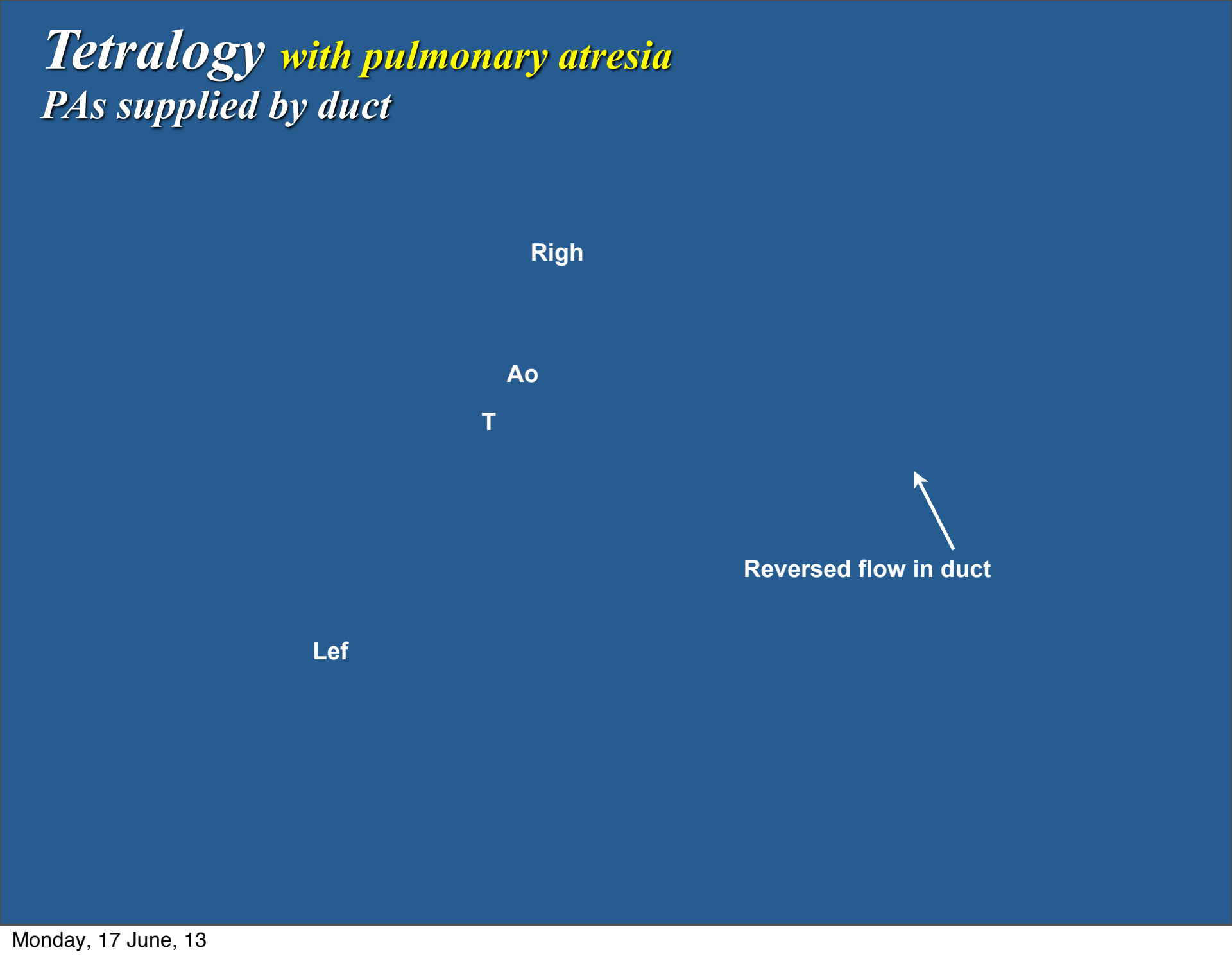
Tetralogy (PA VSD) and MAPCAs



Seale, Ho, Shinebourne, Carvalho. Cardiol Young 2009

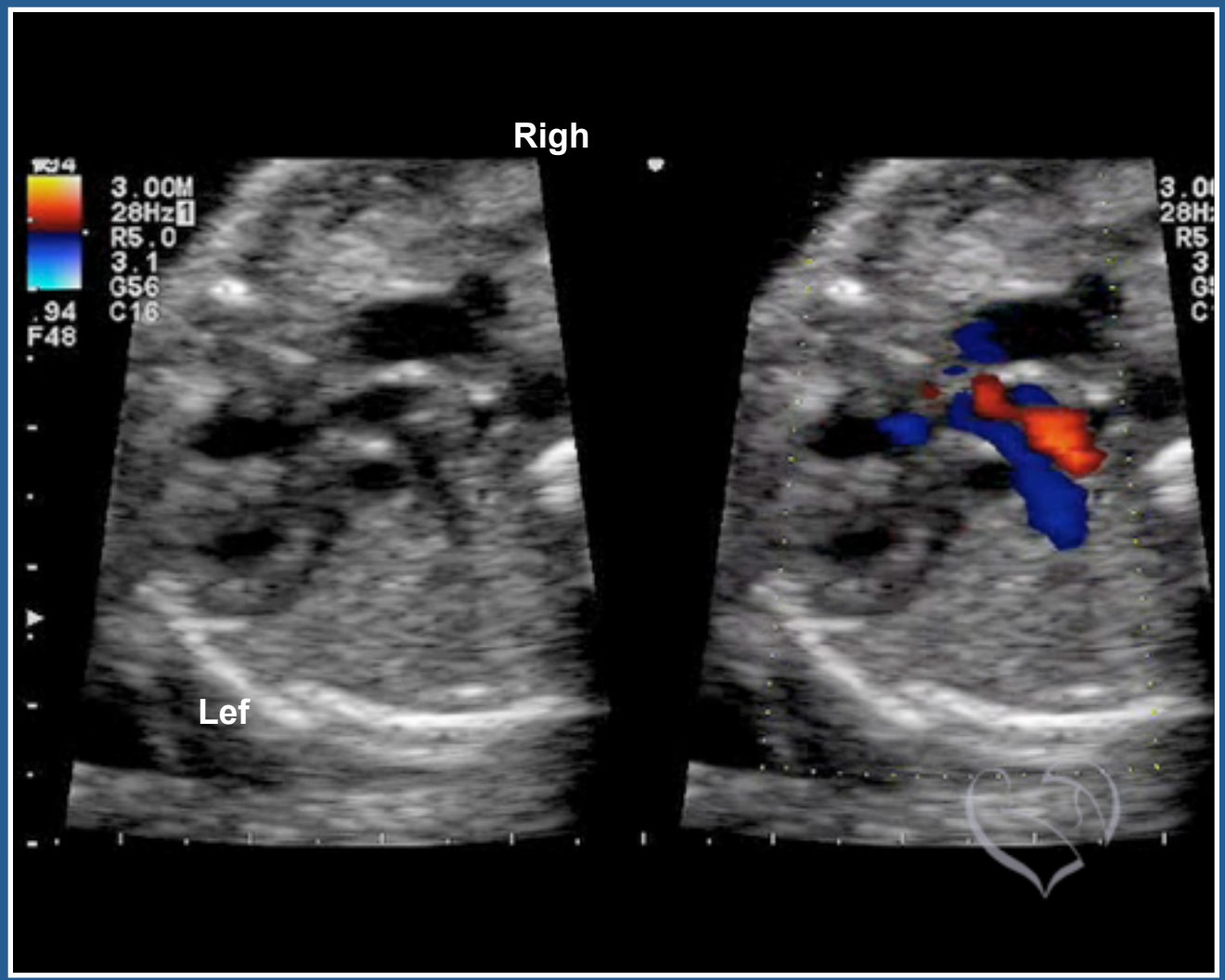
Tetralogy with pulmonary atresia

PAs supplied by duct

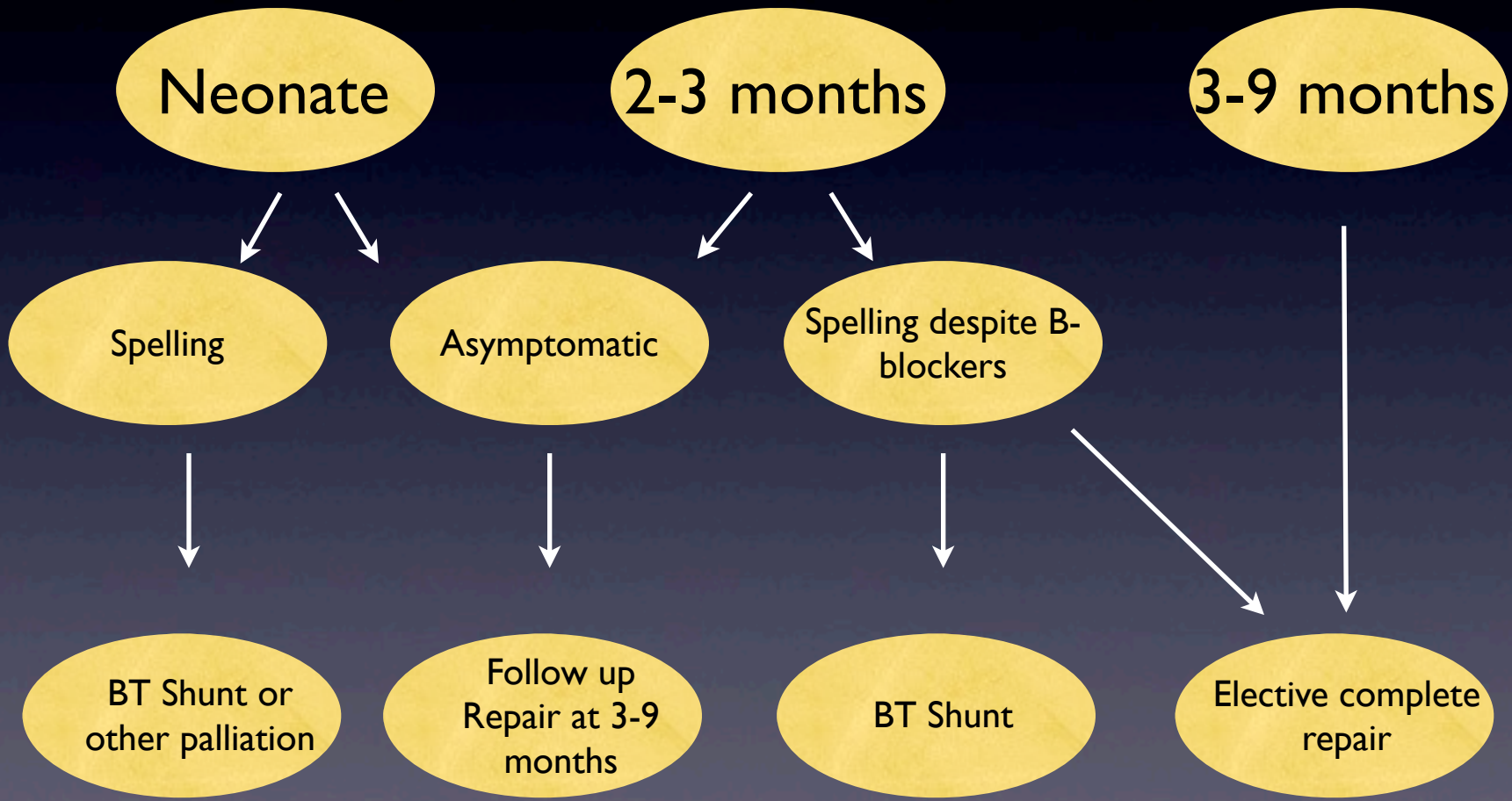


Tetralogy with pulmonary atresia

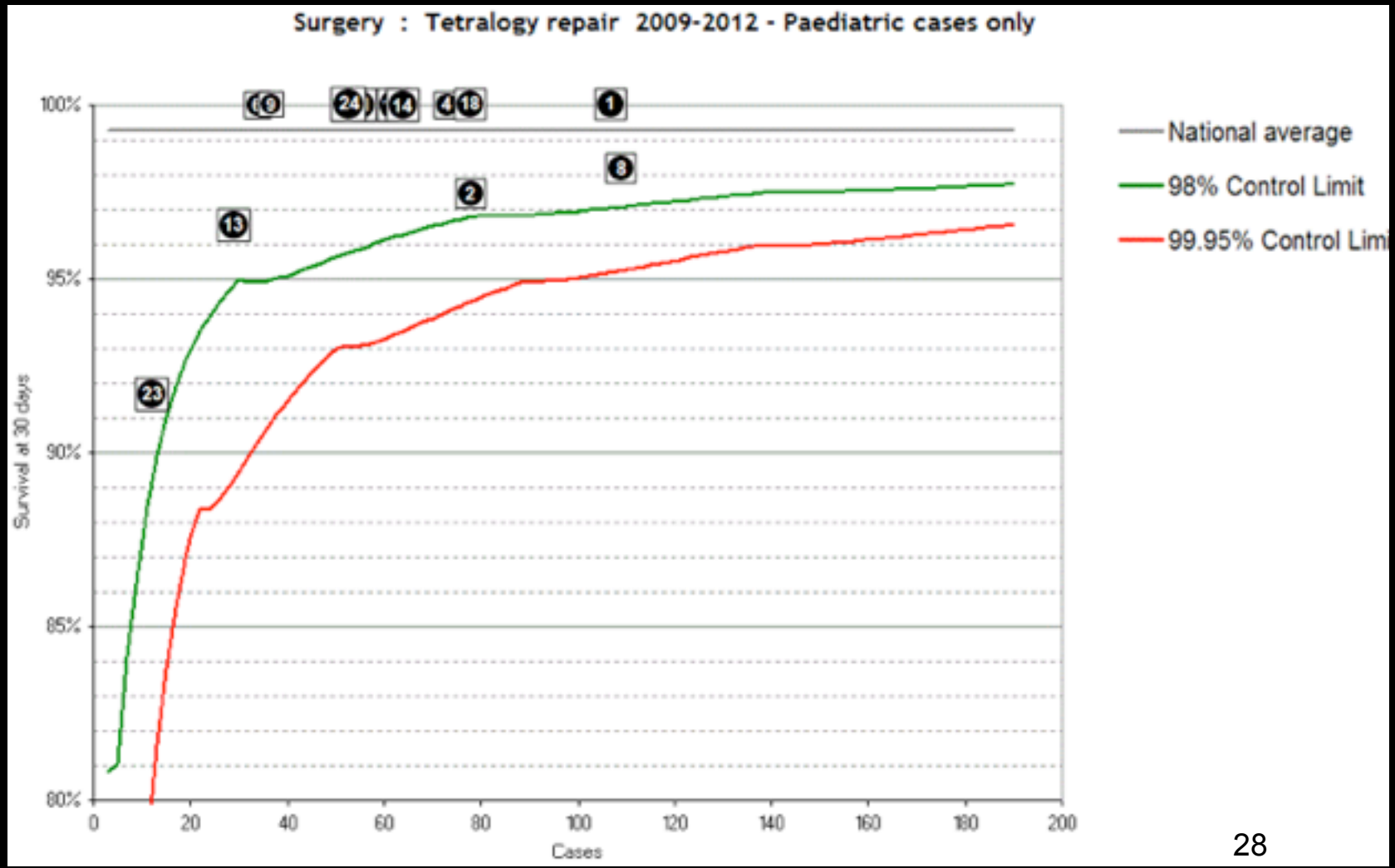
PAs supplied by duct



Regular Tetralogy of Fallot



Repair of Tetralogy of Fallot Today



Conclusion
Optimal management of tetralogy of Fallot

- Strategy should be tailored to the individual patient
- We must preserve the Right Ventricle
- Preserve the pulmonary valve? or Early pulmonary valve implantation?

Fetal service
Diagnostics, counselling, intervention

Neonatal service
Diagnostics, intervention, surgery

Paediatric service
Diagnostics, monitoring; further intervention

Adolescent service
Counselling; education

Transition

Adult Congenital Service
Diagnostics, re-intervention, pregnancy, advanced disease

Congenital heart services at the Trust

Royal Brompton & Harefield NHS
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Adult ToF patients in 30 years
Current surgical era

- Prenatal diagnosis
- Transatrial repair of the VSD
- Transatrial relief of RVOTobstruction
- Combined transatrial/transpulmonary repair
- when pulmonary valve commissurotomy and/or pulmonary trunk enlargement required
- Aim:
- preservation of pulmonary valve/annulus integrity
- Reduced RV scarring, better RV function

but more RVOTO? Is RV restriction good or bad?

L. Boni et al. European Journal of Cardio-thoracic Surgery 35 (2009) 885—890