
Jeudi 3 octobre 2019
10h00-17h30
M3C-Necker
Cardiologie Congénitale et Pédiatrique
Bâtiment Laennec, 4ème étage
149, rue de Sèvres
75015 Paris
M3C Academy
Tétralogie de Fallot

# Simple Tetralogy of Fallot

## Postoperative management made simple (…in 20 minutes)

Stephane LE BEL Hôpital Timone-Enfant

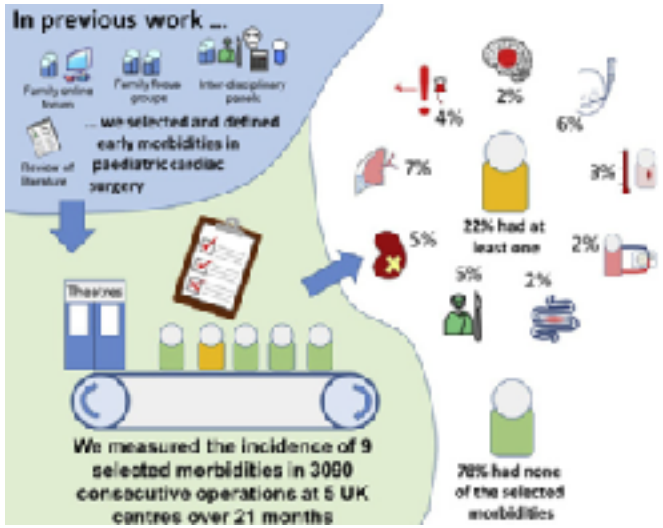
- The severly cyanosed neonate/infant
  - Modified Blalock-Taussig-Thomas shunt
  - RVOT stenting procedure
  - Right ventricule to pumonary artery connection
  - Complete neonatal surgical repair
- Complete surgical repair
- RV-PA conduit
- Melody procedure

# The Anesthetist and Intensivist task
# Risk management

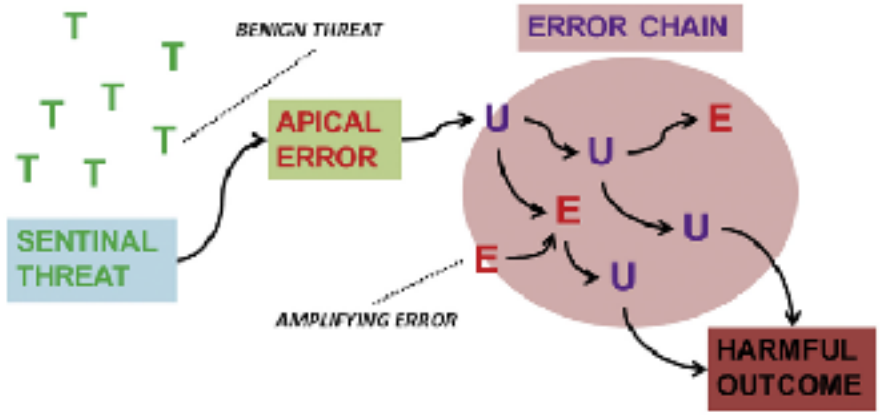
**Incidence and risk factors for important early morbidities associated with pediatric cardiac surgery in a UK population** Brown et al The Journal of Thoracic and Cardiovascular Surgery • ■ 2019

## Odds ratio of Morbidity

- Neonates 5,26 (3,90 - 7,09) p< 0,001
- Complexity 2,14 (1,67 – 3,12) p<0,001
- Bypass > 90’ 2,28 (1,67 – 3,12) p<0,001
- Ilness severity 1,52 (1,16 – 2,00) p<0,01




**NASA Model of “Threat and Error” in Pediatric Cardiac Surgery: Patterns of Error Chains** HICKEY ET AL Ann Thorac Surg 2016




- Sentinal Threat: Anatomy – Comobidities...
- Apical Errors: In the operative theater.
- Additional error chain: In PICU

# Pulmonary to Systemic Anastomosis

JAMA®

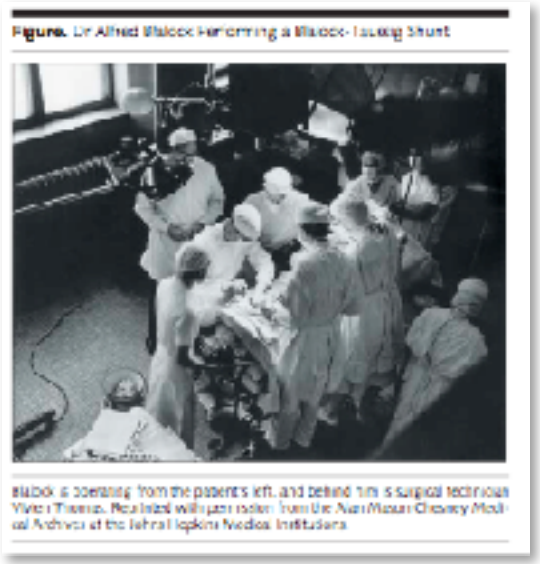
Online article and related content current as of May 5, 2010.

**The Blalock-Taussig-Thomas Collaboration: A Model for Medical Progress**

Anne M. Murphy; Duke E. Cameron

*JAMA*. 2008;300(3):328-330 (doi:10.1001/jama.300.3.328)

http://jama.ama-assn.org/cgi/content/full/300/3/328



THE SURGICAL TREATMENT OF MALFORMATIONS OF THE HEART

IN WHICH THERE IS PULMONARY STENOSIS OR PULMONARY ATRESIA

ALFRED BLALOCK, M.D.

AND

HELEN B. TAUSSIG, M.D.

BALTIMORE

ANESTHESIOLOGY

The Journal of

THE AMERICAN SOCIETY OF ANESTHESIOLOGISTS, INC.

Volume 7 SEPTEMBER, 1946 Number 5

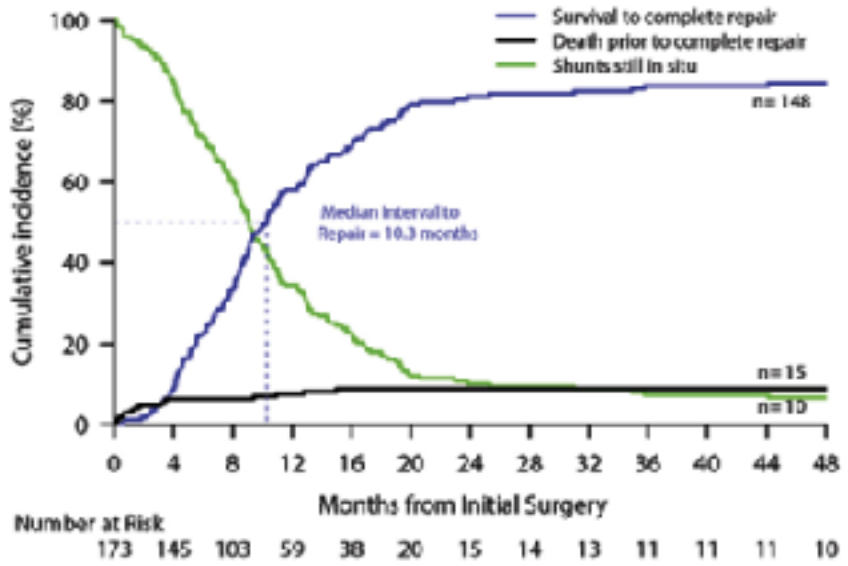
ANESTHESIA IN THE SURGICAL TREATMENT OF CONGENITAL PULMONIC STENOSIS * †

M. H. HARMEL, M.D., AND AUSTIN LAMONT, M.D.

*Baltimore, Md.*

# « It is never just a BT shunt »

**Determinants of Adverse Outcomes After Systemic-To-Pulmonary Shunts in Biventricular Circulation** (Ann Thorac Surg 2017;104:1365–70)




*Results.* [redacted] for the initial shunt procedure. [redacted]. Overall, 86% of patients progressed to corrective surgery. Acute

- 173 children
  - ➢Median age 22 days
  - ➢Median weight 3,2 kg
- Acute Events on 41 patients
  - ➢Chest opening (n=30)
  - ➢Shunt Thrombosis (n=16)
  - ➢Pulmonary overcirculation (n=17)
  - ➢Death (n=6)

# Primary Issues with BT shunt

1. **Volume loading of the heart**
   - Fluids - Transfusion
   - Valvular regurgitation
   - Subendocardial ischemia
2. **Coronary steal**
   - Shunt size - lenght – anastomosis – Pulmonary vs Systemic resistances
   - Low diastolic pressure (PAd<30mmHg)
   - Subendocardial ischemia
3. **Shunt thombosis**
   - Platelets (any pro-coagulant factor)
   - Hemoglobin > 15 g/dl
   - Pulmonary Arterial Hypertension
   - Low Cardiac output
   - Technical issues

# How to deal quickly with a complex physiology?

- SpO2 < 75% vs > 85%
- NIRS $_{\text{cerebral/renal}}$ ratio < 1 vs >1
- ECG (ischemia)
- Invasive Arterial pressure (PAd)
- Arterial gas test (Hb; PaO2; Lactates)
- Echocardiography
- Pulmonary echography
- Cath lab / Tomodensitometry

- **Hypotension and desaturation**
  - ➢Hypovolemia (real or functional) – anemia
  - ➢High pulmonary and low systemic vascular resistance
  - ➢Shunt occlusion/thrombosis with hypoxemic heart failure
  - ➢Pulmonary desease with low pulmonary vein saturation
- **Hypotension and high saturation**
  - ➢High cardiac output with vasodilatation
  - ➢Pulmonary overcirculation with systemic vasoconstirction
- **Normal PAm / PAd and low SpO2**
  - ➢Technical Issues
  - ➢High PVR - PAHT
  - ➢High O2 consumption

# RVOT and Ductal stenting are high-risk procedures

Procedural characteristics and adverse events in diagnostic and interventional catheterisations in paediatric and adult CHD: initial report from the IMPACT Registry

*Cardiology in the Young* (2016)

**CRISP: Catheterization RISk Score for Pediatrics:** A Report from the Congenital Cardiac Interventional Study Consortium (CCISC)

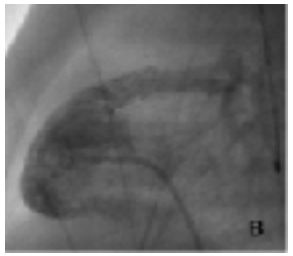
**Nykanen et al.** Catheterization and Cardiovascular Interventions 87:302–309 (2016)

## Neonatal interventional catheterism

- All cause mortality 12%
  - ➢1st primary cause: Cardiac 65,1%
  - ➢2nd primary cause: Pulmonary (16,6%)
- Adverse Events 30,2%
  - Major AE 4,2%

| Assigned Points | 0 | 1 | 2 |
|---|---|---|---|
| Patient Status/Timing ($X_1$) | Elective | Emergent/Urgent | Post-operative |
| Age ($X_2$) | > 1 year | 30 days–1 year | < 30 days |
| Weight ($X_3$) | > 10 kg | 2.5–10 kg | < 2.5 kg |
| Inotropic Support ($X_4$) | None | Yes–Stable | Yes–Unstable or ECMO |
| Respiratory Status ($X_5$) | Own Airway | Stable on ventilator or known difficult/unusual airway | Respiratory failure on mechanical ventilation |
| Systemic Illness/failure ($X_6$) | none | Medically controlled or 1 organ system failure | Uncontrolled or > 1 organ system failure |
| ASA Score ($X_7$) | 1 or 2 | 3 | 4 or 5 |
| Physiologic Category ($X_8$) | Category 1 | Category 2 | Category 3 |
| Pre-Catheterization Diagnosis ($X_9$) | Category 1 | Category 2 | Category 3 |
| Procedure Risk Category ($X_{10}$) | Category 1 | Category 2 | Category 3 |

- **A two month infant, with T4F, severly desaturated, avlocardyl, two fluid challenge, non Invasive ventilatory support (CPAP), schedulled for RVOT stenting procedure**
- **CRISP=9 ➔ Serious Adverse Events = 8-10%**



# RVOT / Ductal stenting

## Non specific issues

- Poor patient access
- Poor lighting
- Radiation exposure
- Offsite location
- Low ambient temperature
- Frequent flushings
- Positioning issues
- Bleeding
- Vascular trauma/thrombosis
- Dysrythmia
- Vascular / cardiac chamber perforation
- Stroke /air embolism

## Specific issues

- Patent ductus arteriosus?
  - ➢ Alprostadil induced morbidities
- Complex DA anatomy
- Guiding and stent positioning
  - ➢ Fluid chalenge
  - ➢ Esmolol
  - ➢ Vasopressor
- Emergent surgery
- Greater need for reintervention

## Comparison Between Patent Ductus Arteriosus Stent and Modified Blalock-Taussig Shunt as Palliation for Infants With Ductal-Dependent Pulmonary Blood Flow

### Insights From the Congenital Catheterization Research Collaborative

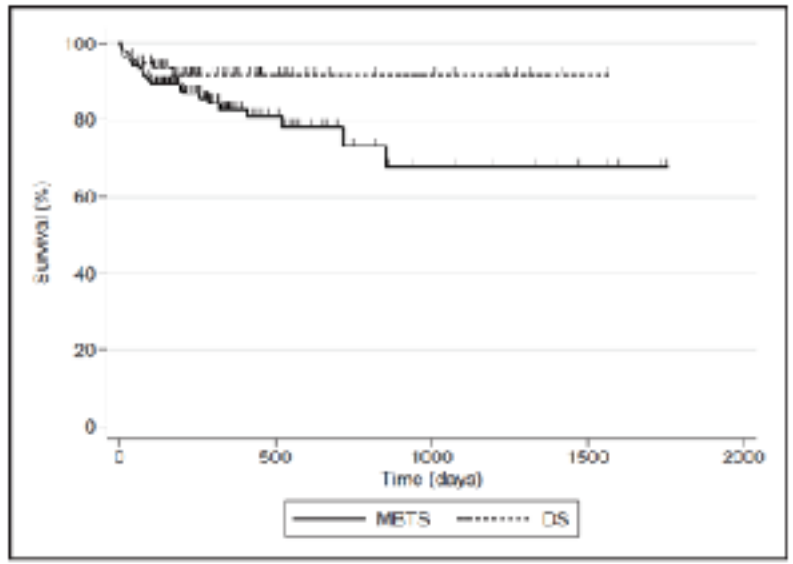
*Circulation.* 2018;137:589–601

These findings support patent ductus arteriosus stent placement as a preferable alternative palliative strategy to Blalock-Taussig shunt placement in select patients with ductal-dependent pulmonary blood flow, where this procedure can be performed safely and effectively.

## Duct Stenting Versus Modified Blalock-Taussig Shunt in Neonates With Duct-Dependent Pulmonary Blood Flow

### Associations With Clinical Outcomes in a Multicenter National Study

*Circulation.* 2018;137:581–588.




| | MBTS, n (%) | DS, n (%) | Comparison *P* Value |
|---|---|---|---|
| Length of stay in hospital, median (IQR), d | 21 (14–31) | 14 (7–22) | <0.001 |
| Length of stay in intensive care unit, median (IQR), d | 7 (4–15) | 2 (0–6) | <0.001 |
| Length of stay on ventilation, median (IQR), d | 4 (2–8) | 1 (0–4) | <0.001 |

In favor of arteriosus stenting:

- Fewer procedural complication
- Shorter ICU LOS
- Less diuretics
- No difference in the hazards of a composite outcome (death + unplanned intervention to treat cyanosis)

# Stenting of the right ventricular outflow tract

Oliver Stumper, Bharat Ramchandani, Patrick Noonan, Chetan Mehta, Vinay Bhole, Zdenka Reinhardt, Rami Dhillon, Paul A Miller, Joseph V de Giovanni

Stumper O, *et al*. *Heart* 2013;**0**:1–6.

**Table 1** Clinical factors and associated anatomic lesions

| | n |
|---|---|
| Clinical factors | |
| Weight <3 kg | 17 |
| Saturations <75% | 25 |
| Recent spell | 26 |
| Prostaglandin infusion | 12 |
| Severe genetic syndrome | 14 |
| Recent necrotising enterocolitis | 4 |
| Recent bronchiolitis/severe CLD | 6 |
| Tracheostomy/tracheal stenosis | 4 |
| Anatomical factors | |
| Hypoplastic pulmonary arteries | 19 |
| MAPCAs | 5 |
| AVSD | 8 |
| Tricuspid atresia | 1 |
| Isomerism | 3 |
| DORV | 9 |
| Left SVC to left atrium | 4 |
| Coronary anomaly | 3 |

AVSD, atrioventricular septal defect; CLD, chronic lung disease; DORV, double outlet right ventricle; MAPCA, major aortopulmonary collateral artery; SVC, superior vena cava.

- 52 patients
- 1 per-procedure death
- 1 emergent surgery
- 2 BTS shunt for inadequate oxygenation.
- 16 re-catheterization
  - ➢Baloon angioplasty
  - ➢Re-stent procedure

# Neonatal right ventricle to pulmonary connection as a palliative procedure for pulmonary atresia with ventricular septal defect or severe tetralogy of Fallot[†]

Sébastien Gerelli[a], Mathieu van Steenberghe[a], Bari Murtuza[a], Mirela Bojan[a], Ekoué Diana Harding[a], Damien Bonnet[c], Pascal R. Vouhé[a] and Olivier Raisky[a,*]

European Journal of Cardio-Thoracic Surgery 45 (2014) 278–288

APSO: classification

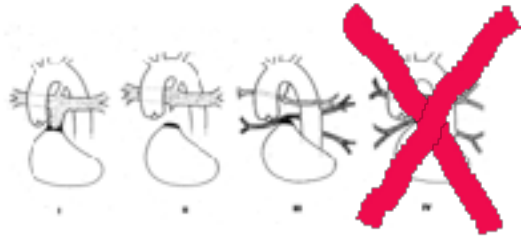


**Table 2:** Operative and postoperative characteristics after RV to PA connection

| Variable | PA/VSD I ($n$ = 24) | PA/VSD II ($n$ = 14) | PA/VSD III ($n$ = 7) | TOF ($n$ = 12) | Total ($n$ = 57) |
|---|---|---|---|---|---|
| CPB duration (min) | 69 [26–188] | 83 [45–163] | 77 [38–96] | 54 [34–110] | 64 [26–188] |
| Cross-clamping duration (min) | 26 [13–57] | 29 [16–76] | 30 [18–32] | 23 [14–59] | 25 [13–76] |
| Postoperative characteristics | | | | | |
| Mechanical ventilation (h) | 41 [17–465] | 48 [12–312] | 45 [19–96] | 42 [5–288] | 48 [5–600] |
| Length of ICU stay (days) | 3 [1–35] | 5 [2–18] | 4 [3–14] | 6 [1–16] | 4 [1–35] |
| Arterial saturation (%) | 90 [85–95] | 95 [92–95] | 94 [82.5–95] | 92 [89–95] | 92 [70–99] |
| Delayed sternal closure | 4 (16.7) | 3 (21.4) | 1 (14.3) | 8 (66.7) | 16 (28) |
| Ventricular dysfunction | 5 (20.8) | 3 (21.4) | – | 3 (25) | 11 (19.3) |
| Excessive pulmonary blood flow | 2 (8.3) | 4 (28.6) | – | 2 (16.7) | 8 (14) |
| Restrictive pulmonary blood flow | 3 (12.5) | 1 (7.1) | 1 (14.3) | | 5 (9) |
| BT shunt | 3 (12.5) | 1 (7.1) | – | – | 4 (7) |
| Enlargement RV to PA | – | – | 1 (14.3) | – | 1 (2) |
| Sepsis | 3 (12.5) | 2 (14.3) | – | – | 5 (8.8) |
| Prolonged pleuro-pericardic effusion | 2 (8.3) | 1 (7.1) | – | 2 (16.7) | 5 (8.8) |
| Phrenic paralysis | – | 1 (7.1) | – | – | 1 (1.8) |
| Kidney failure requiring dialysis | 1 (4.2) | | | | 1 (1.7) |
| Hospital death | 1 (4.2) | – | – | 1 (8.3) | 2 (3.5) |

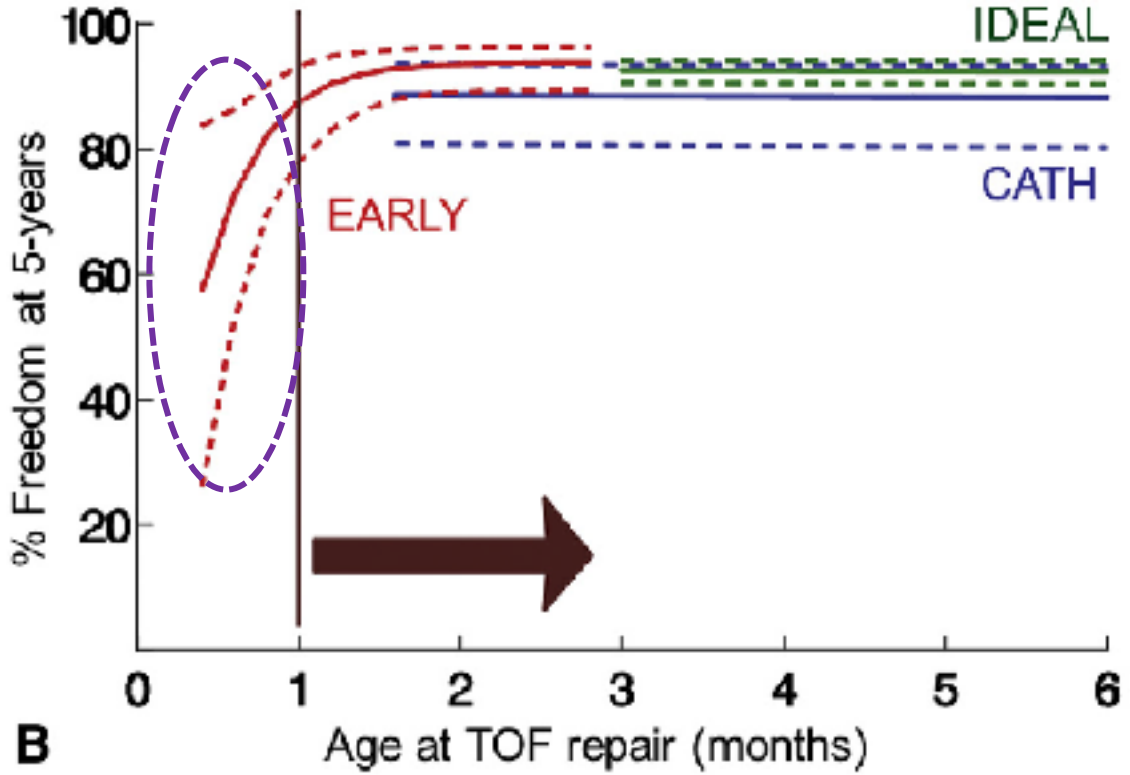
Data are shown as medians and range (minus, major) or as numbers and percentages.
CPB: cardiopulmonary bypass; ICU: intensive care unit; BT: Blalock–Taussig

# Young infants with severe tetralogy of Fallot: Early primary surgery versus transcatheter palliation

J Thorac Cardiovasc Surg 2017;154:1692-1700

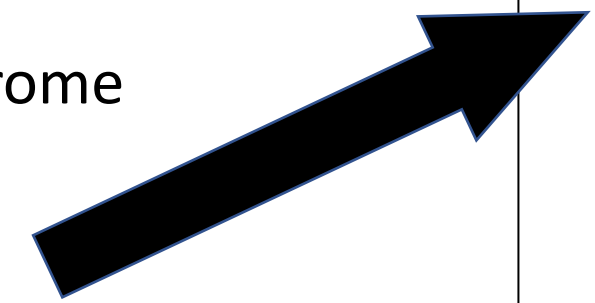
| Variable | IDEAL (N = 383) Median (range) or N (%) | EARLY (N = 42) Median (range) or N (%) | CATH (N = 28) Median (range) or N (%) | P* |
|---|---|---|---|---|
| Age at first intervention (d) | 189 (92-6527) | 50 (5-90) | 13 (1-305) | <.001 |
| Age at primary repair (mo) | 6.3 (3.1-184) | 1.6 (0.3-2.7) | 4.5 (1.4-13) | <.001 |
| BSA ($m^2$) at repair | 0.4 (0.23-1.9) | 0.27 (0.17-0.39) | 0.37 (0.24-0.78) | <.001 |
| Male | 214 (56%) | 26 (62%) | 13 (46%) | .15 |
| Death | 11 (1%) | 1 (2%) | 1 (4%) | .49 |

| Baseline morphology | Mean (SD) or N (%) | Mean (SD) or N (%) | Mean (SD) or N (%) | |
|---|---|---|---|---|
| Operative summary | | | | |
| RVSP after initial intervention | 46 ± 11 | 44 ± 9 | 57 ± 14 | .058 |
| CPB (min) | 113 ± 43 | 104 ± 33 | 146 ± 59 | .013 |
| Ischemic time (min) | 72 ± 24 | 75 ± 26 | 88 ± 32 | .57 |
| Transannular patch | 103 (27%) | 16 (38%) | 17 (61%) | .088 |
| Pulmonary valvotomy | 237 (62%) | 16 (38%) | 5 (18%) | .11 |
| Conduit | 12 (3%) | 8 (19%) | 2 (7%) | .50 |
| Final PV probe diameter (mm) | 10.4 ± 2 | 9.6 ± 1 | 10.1 ± 0.6 | .051 |
| Final RVSP (mm Hg) | 46 ± 11 | 44 ± 9 | 46 ± 11 | .46 |
| Final gradient (mm Hg) | 20 ± 9 | 18 ± 15 | 14 ± 5 | .06 |
| Surgical reoperations† | 29 (8%) | 7 (17%) | 3 (11%) | n/a |
| Catheter reintervention† | 39 (10%) | 11 (26%) | 11 (39%) | n/a |



# TOF complete surgical repair

- Junctional Ectopic Tachycardia
- Postoperatie bleeding
- Low Cardiac Output Syndrome
  - ➢Tamponade
  - ➢**RV Systolic dysfunction**
  - ➢**RV Diastolic dysfunction**
- Residual lesions
  - ➢Branch PA stenosis
  - ➢Residual RVOT obstruction
  - ➢Residual/new VSD
- Ileus / Poor enteral feeding



- Right Ventricular Dysfunction
  - ➢RV muscle hypertrophy limits cardioplegia efficiency
  - ➢RVOT résection and related arythmie / heart block
  - ➢RV hypetrophy and diastolic dysfunction

**Transesophageal Echocardiography in Tetralogy of Fallot**

*Seminars in Cardiothoracic and Vascular Anesthesia 16(2)*

- Residual VDS>4-5mm
- RVOT velocities > 3m/s
- Pulmonary regurgitation

Cerebral and Somatic Oxygen Saturations After Repair of Tetralogy of Fallot: Effects of Extubation on Regional Blood Flow

Ann Thorac Surg 2013;95:682–6

# Postoperative clinical pathway

**Day 0**

- Volemia
  - ➢RAP ≈ 5-10 mmHG (15mm Hg)
- HR<150/mn
  - ➢T° control (SIRS)
  - ➢Analgesia - Dexmedetomidine
  - ➢SuMg+; Adénosine; amiodarone
- Normal Blood Pressure
  - ➢Vasoplegia (SIRS – volemia – vasopressor)
  - ➢LCOS (milrinone)
  - ➢PFO++

**Early Extubation After Repair of Tetralogy of Fallot and the Fontan Procedure: An Analysis of The Society of Thoracic Surgeons Congenital Heart Surgery Database**

Ann Thorac Surg 2016;102:850–8

- 1153 TOF repairs (92 centers)
- Mortality 0,13%
- Early Extubation (< H+6h) 31%
- Median ventilation duration 20,9 h
- Early Extubation associated factors:
  - ➢Higher weight
  - ➢Lesser skin incision to skin closure time.
  - ➢Lesser aortic cross clamping time

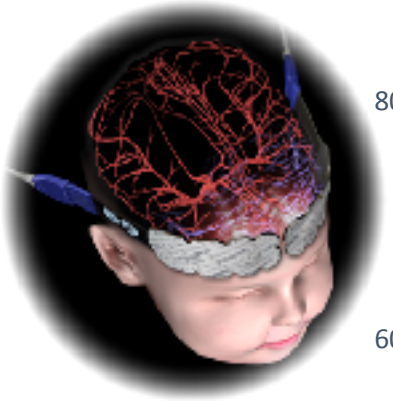
# Right Ventricule to Pulmonary Conduit

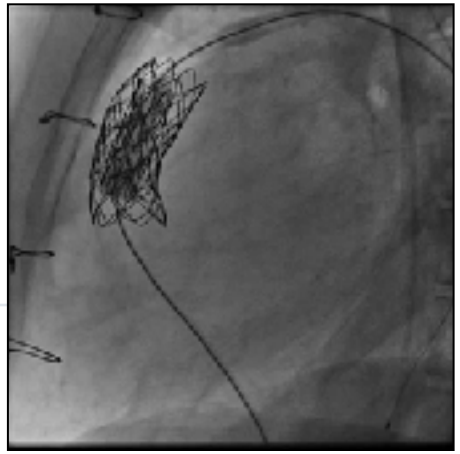
- Redo surgery
- Low morbidity related to pre-CPB dissection time and complication (bleeding)
- Fast track pathway
- Short ICU stay

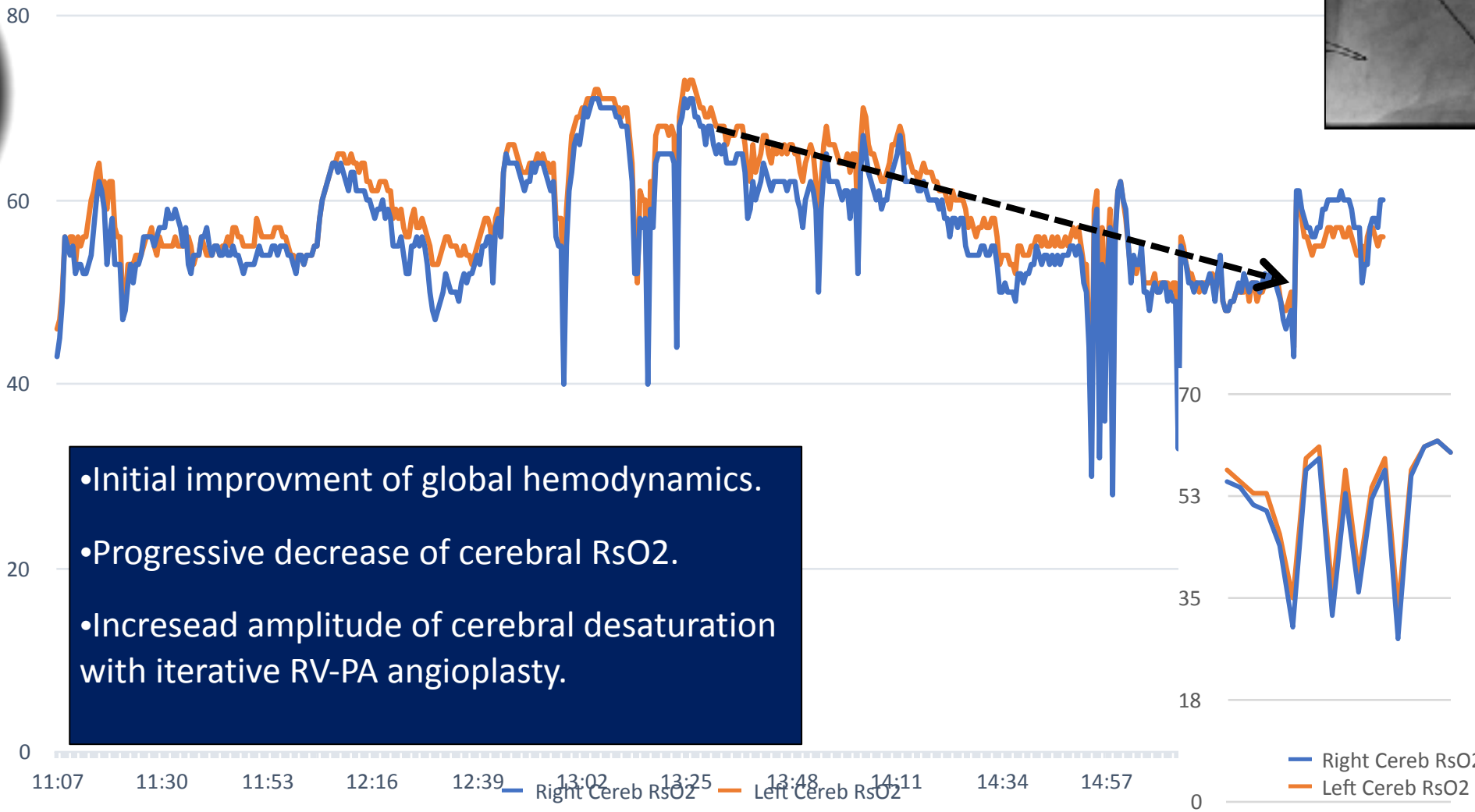
# Percutaneous Pulmonary Valvulation

- Patient comorbidities:
  - ➢Right ventricular failure
  - ➢Distal pulmonary artery stenosis
- General anesthesia
- Prolonged procedure
  - Pressure injury
  - Plexus injury
- Hemoptysis
- ICU / CCU admission unfrequent

# Iterative balloon inflations led to an impaired hemodynamic status with poor tolerance in a 14 years old girl with repaired TOF and right ventricular systolic failure








- Initial improvment of global hemodynamics.
- Progressive decrease of cerebral RsO2.
- Incresead amplitude of cerebral desaturation with iterative RV-PA angioplasty.

# Conclusion

- Mortality and morbidity belong to the cohort of profundly desatured neonates.
- Surgical or percutaneous intervention are high risk procedure in this context.
- Anesthesiologist and Intensivist are the ones confronted with morbidities occurrence and treatment.
- Well known neonatal palliative procedure vs new percutaneous technique?
- How can we improve neonatal acute care?
- How can we prevent or decrease the occurrence of postoperative adverse events?