Transcatheter Intervention for Preoperative Symptomatic TOF

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Yonsei Pediatric Cardiology

Tetralogy of Fallot

✓ About 6~8% of CHD

- Elective surgical correction usually 2-3~6mo

✓ Young/small infants with high risk for surgical repair & significant Sx (cyanosis and/or hypoxic spell)

Treatment options

- Early total repair
- Palliation (staged repair)
  1. systemic-pulmonary shunt / palliative RVOT enlargement
  2. interventional palliation
     : RVOT balloon dilatation / RVOT stenting
     (PDA stenting / cutting balloon dilation?)

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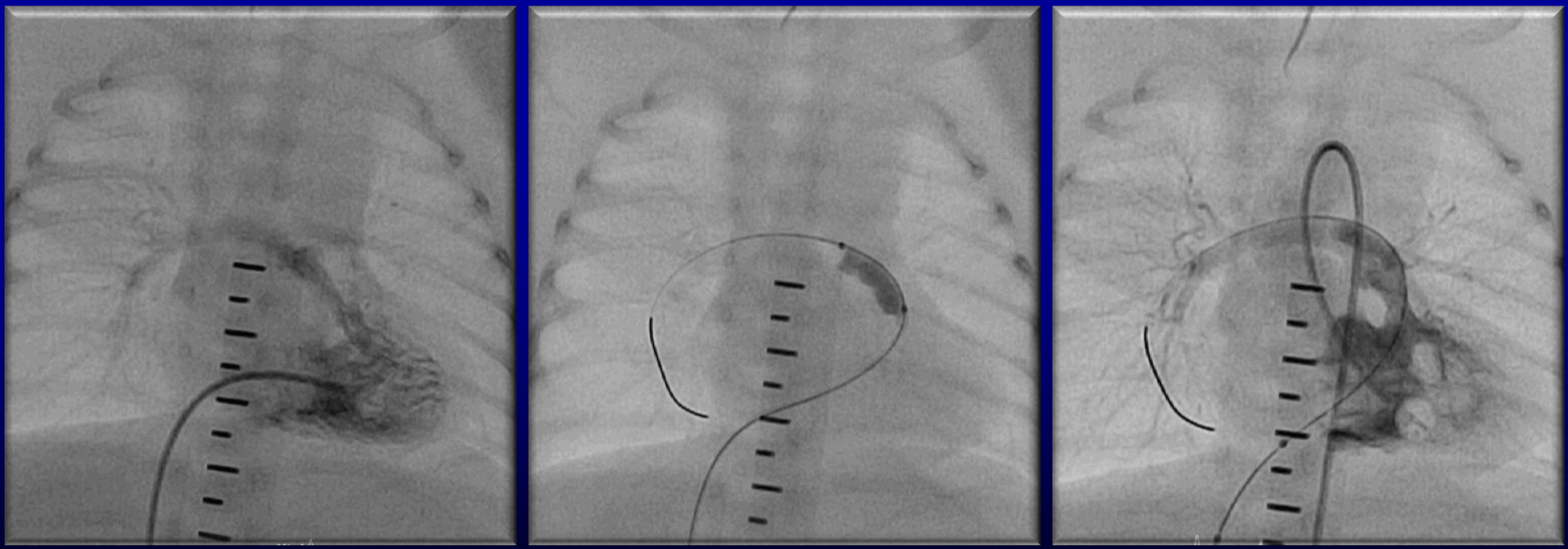
Balloon dilation of RVOT

- Efficacy & safety has been reported since 80's

Qureshi SA. Br Heart J 1988, Boucek MM. Am Heart J 1988, Rao PS. Am Heart J 1988

KJR F/20days

- Down SD, DORV with subarterial VSD, severe PS
- Cyanosis with periodic worsening



Balloon dilation of RVOT

- Indications -

✓ **Young infant : 0~2(or 3)mo – not amenable to corrective surgery at the time of presentation**

- **Symptoms related to cyanosis:**

  SaO2↓ <75~80% / Episodes of hypoxic spell

- **Small/diminutive PAs, coronary anomalies (crossing RVOT), AVSD, or other conditions with high risk for definitive surgery**

- **Case selection**

  - ***Predominant valvular stenosis – best candidate***

    ass infund/annular narrowing: usually not regareded as contralx

    * **predominant infundibular obstruction**

    - not likely efficacious / effect not last long enough
    - more risky (eq. procedure induced spell)

  - ***should be cautious for pts likely cause pulm flooding by BPV***

Equipment – Balloons/Sheaths/Wires

✓ Balloon catheters

- Tyshak valvuloplasty catheters – most commonly used
(quick inflation & deflation / lower profile)

- usually 20mm length

- coronary balloons may be initially used for a tight valve
→ staged balloon dilatation to intended diameter

- higher pressure balloons – (COEfficient, Z-med line)
not preferred, may have potential role for dysplastic valves

✓ Introducer

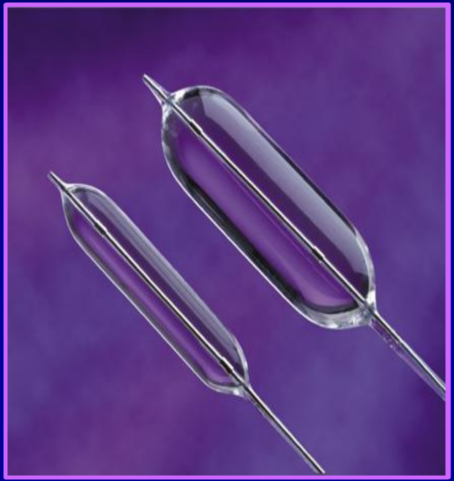
- 4Fr introducer for low pressure balloons (Tyshak ~8mm)
- higher pressure balloons – 1-2Fr larger sheath

✓ Wires

- Tyshak II : .021" wire (~8mm balloon)
- Tyshak mini : .014" coronary wires
- COEfficient : Roadrunner (.018")

✓ Frequently used Balloon Catheters
mostly from NuMed company






Tyshak II
4Fr/4~8mm
burst pr :
4~6atm/4~8mm
GW: .021"

Tyshak mini
3Fr/4~8mm
burst pr :
4~6atm/4~8mm
GW: .014"

Z-med II
6~7Fr/5~8mm
burst pr :
15atm/4~8mm
GW: .025"
.035"/8mm

COEfficient
4~6Fr/4~8mm
burst pr :
10~13atm/4~8mm
GW: .018"

Techniques

✓ **Pre-procedural**

- adequate hydration
- preparation for spell Tx / administration of propranolol
- anesthetic / surgical back-up

✓ **Procedure**

- adequate sedation or general anesthesia
- SatO2 monitoring
- access: usually 1 femoral vein + 1 femoral artery (BP / blood gas)
- routine cath with biplane RV angio (hand / machine injection)
- 4~5Fr JR (or multipurpose) → deep position of wire (commonly RPA)
- balloon size: 1.2~2.0 x PV annulus/ 2cm long, quick inflation/deflation
- confirming exit angio / SaO2 measurement / checking VS

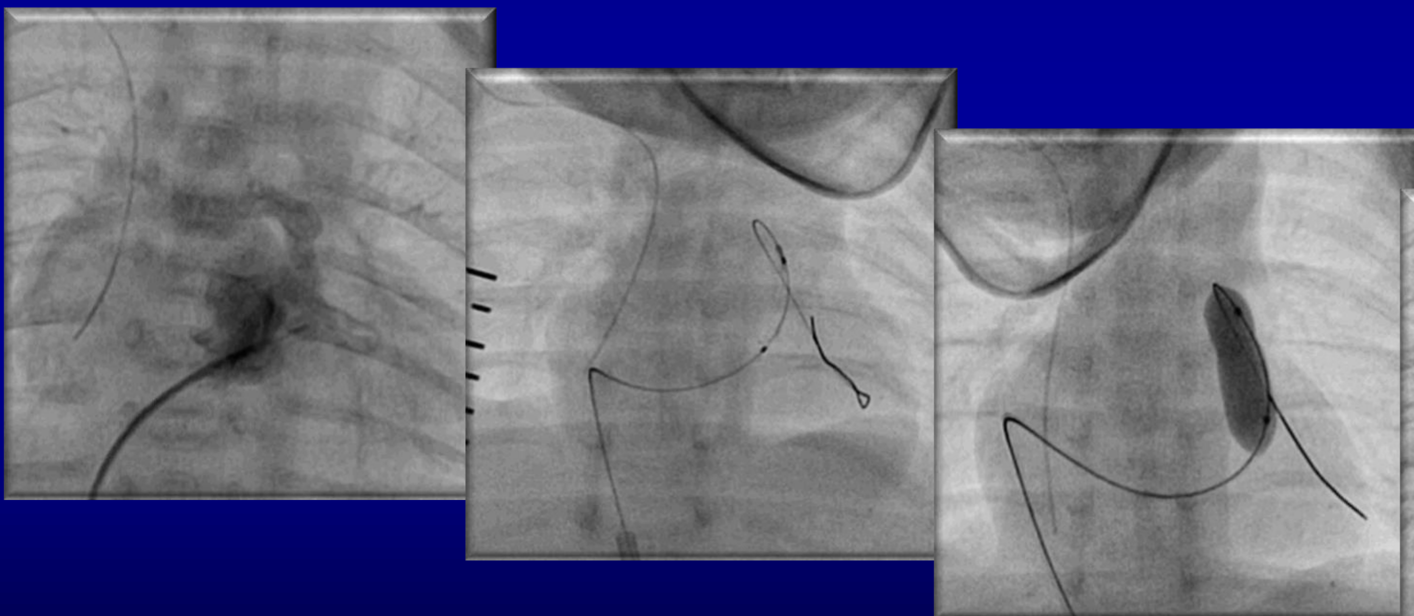
Tips: staged ballooning / high pressure ballooning as needed

✓ **Post-procedural**

- adequate hydration
- monitoring SatO2 / BP / urine output
- β-blocker (propranolol) for infundibular reaction

Case CSW (M/2mo)

- Down SD, Cong megacolon s/p Soave op., FTT
- Systemic infection with desaturation
- Staged ballooning
- 4*15mm coronary balloon → 7*20mm Tyshak II
- Total repair at 1yr of age



Balloon dilation at 2mo of age

F/U at 1yr of age

Outcome

- **Proven efficacy & safety since initial experiences in 1980's**

  Qureshi SA. Br Heart J 1988, Boucek MM. Am Heart J 1988,
  Rao PS. Am Heart J 1988

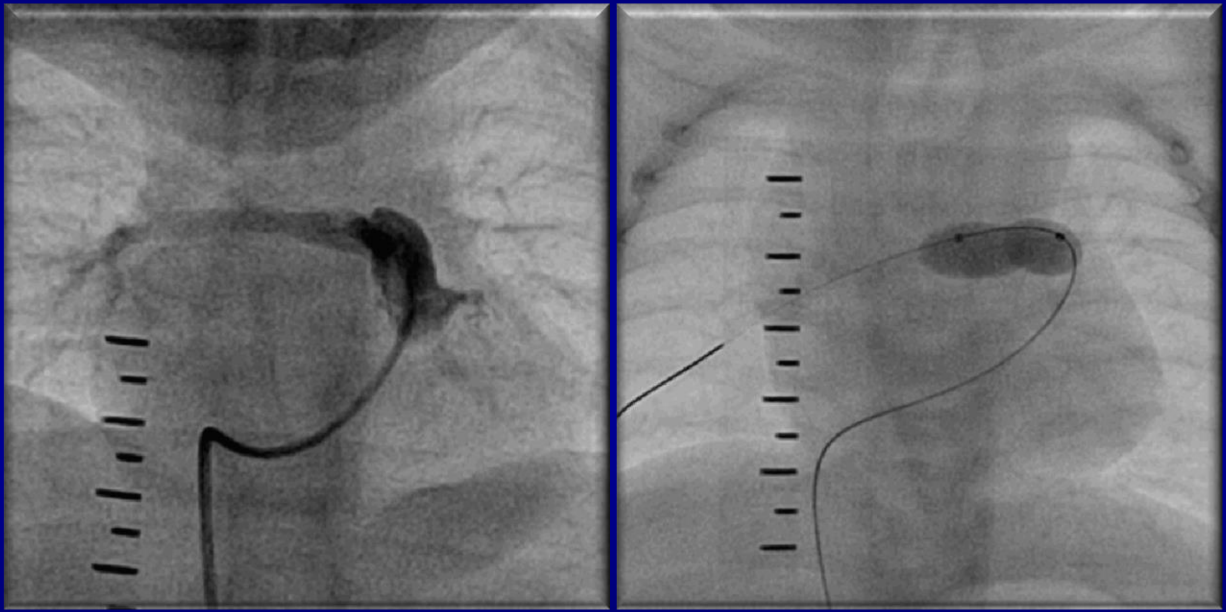
- **SaO_2↑, Hct↓, Promotes PA/ PV annulus growth**

  → ***need of TAP↓*# (30~40% reduction*)***

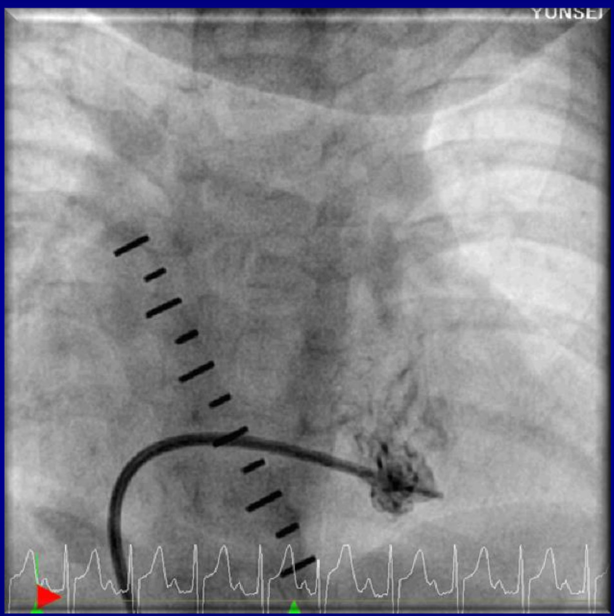
  Parsons JM. Br Heart J 1989, Sluysmans T. Circulation 1995, Kreutzer J. JACC 1996, Godart F. Eur Heart J 1998#, Massoud I1. Cardiol Young 1999, Remadevi KS. Ann Pediatr Cardiol 2008, Cholkraisuwat E. J med Assoc Thai 2010…*

  ↔ *contradictory report: Battistessa SA. Br Heart J 1990, Heusch A. Cardiol Young 1999*

Does PA grow after balloon dilation? Case 1

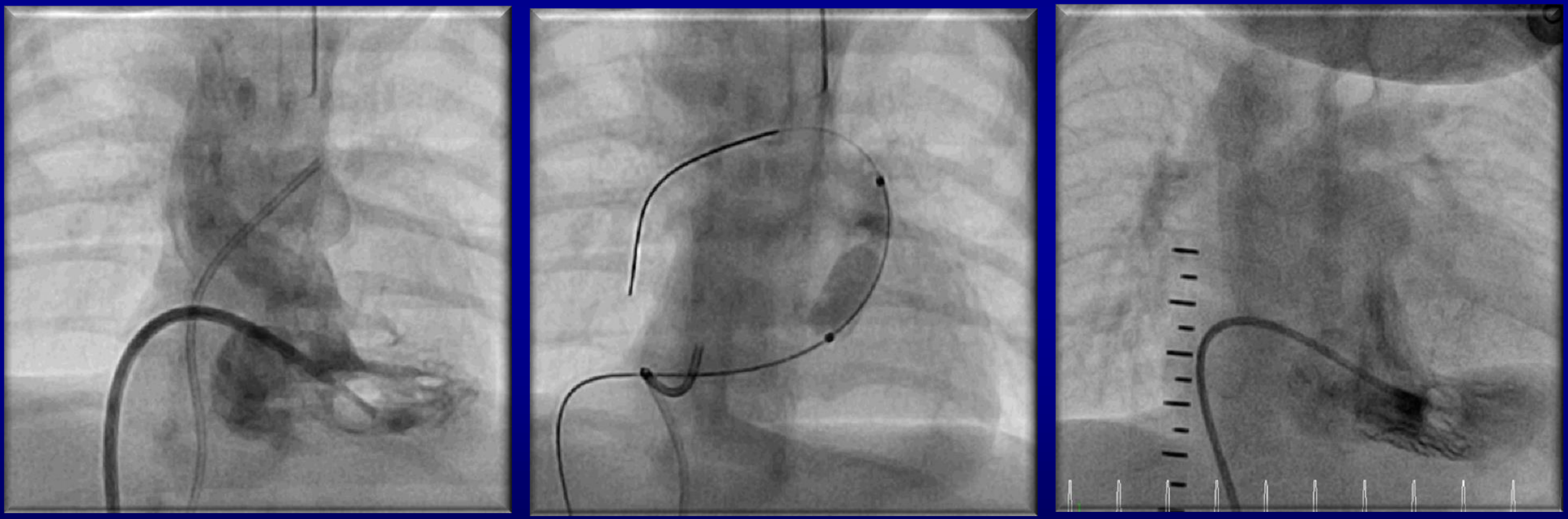


BPV at 2wks of age



F/U at 5mos of age

Does PA grow after balloon dilation? Case 2



BPV at 2days of age

F/U at 5mos of age

Complications

- **Usually reported to be minimal**
- **Some authors reported mod~serious complications**

  - Arab SM. Cardiol Young 1999 : 15 older patients (1.9, 0.5~3yrs) with anoxic spell
  3/15 (20%) proced failure, 2/3 failure – died of severe anoxic spell (2/15, 15%)

  - Remadevi KS. Ann Pediatr Cardiol 2008
  1/17 (6%) RVOT perforation requiring pericardiocentesis → later RVOT aneurysm

  - Heusch A. Cardiol Young 1999
  3/27 (11%) hypoxic spell (recovered), 4/27 (15%) deep vein thrombosis

- **Other problems of BPV in young symptomatic TOF infants**

  - Short lasting efficacy ~20%,
  - Hypoxic spell ~20%
  - Efficacy occ. unpredictable (may be d/t dynamic infundibular obst)
  - Damage (tearing/split) to the leaflet(s) ~50%

  Sluysmans T. Circulation 1995

RVOT Stenting
- Indications -

✓ **Fundamentally and intrinsically, similar to Ix of BPV, however reported cases in the literature mainly targeted slightly different patient group**

- **Severe cyanosis / hypoxic spells**
- **PGE dependent pulmonary circulation**
- **High-risk group for corrective or palliative surgery**
  - **- low weight**
  - **- significant co-morbid conditions**
    prematurity ± ICH / lung disease / genetic disorders..
- **Pulmonary atresia / no effect or non-sustained effect after BPV**

✓ **Common candidates**

- usually unsalvageable pulm valve

- more liberally involve pts with infundibular stenosis

Equipment – Balloons/Sheaths/Wires

✓ **Stents**

- Premounted bare metal stents - most commonly used

  Omnilink (Abbott), Pamaz Genesis (J&J), Express (Boston Scientific), Scuba (Medtronic)…

- Coronary stents (available up to 4.5mm diameter)

  : for very small infants < 3kg

- Hand-crimped stents: delivery sheath > 1-2Fr than balloon

✓ **Delivery sheath**

- 4-5Fr Flexor sheath / 6Fr guiding catheter
- other long sheaths compatible with stent/balloon

✓ **Wires**

- compatible wires with stent/balloon
- .014” wires for coronary stents
- wires for delivery sheath (.035~.038”) can be different from wires for other equipment (.014”~025”)

✓ Available Pre-mounted Stents

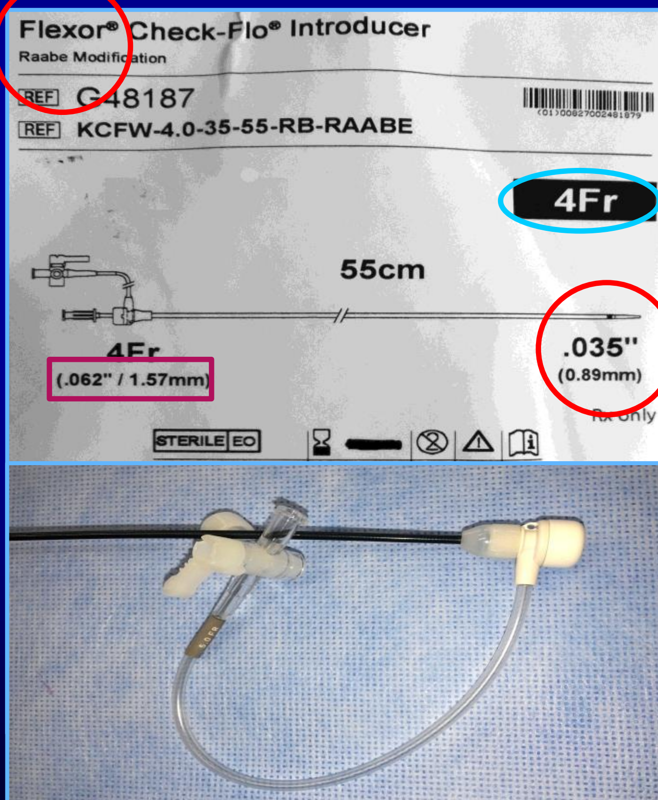
mostly manufactured for iliac, renal, biliary…

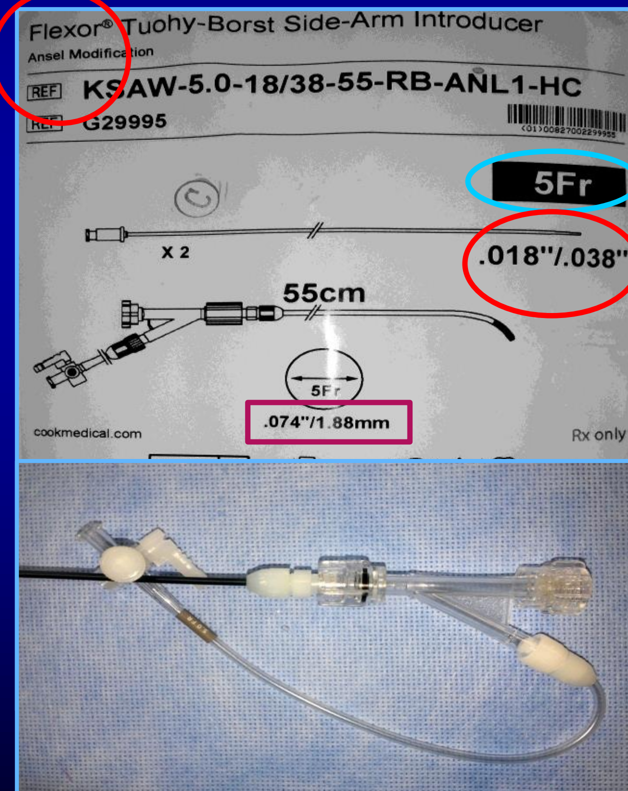
Company	Product	Sheath (W)	Diameter	Length	Mat
Abbott	Omnilink Herculink	6 (.035) 5 (.014)	4~10 4~7	12,16,19,29.. 12,15,18	CoCr CoCr
Boston Scientific	Express Express LD	5-6(.018) 6-7(.035)	4~7	14,15,18,19 17,25,27,37..	SS SS
COOK	Fomula 414 Fomula 418	5(.014) 5-6(.018)	4~7 3~8	12,16,20,24 12,16,20,24,30	SS SS
Cordis	Palmaz Gen Palmaz Blue	6-7(.035) 4-5(.014/8)		11,14,17,23 +10,12,15,18,24	SS SS
Medtronic	Scuba Racer	6(035) 5-6(.014/8)	5~10 4~7	18,30,37.. 12,18	CoCr CoCr
Covidien	Paramount Mini GPS	5-6 (.014-.018)	5~7	14,18,21	SS
Bard	Valeo	6-7(.035)	6~10	18,26,36	SS

** Coronary stents or hand-crimped stents can also be used according to the characteristics of patient and target lesion*

✓ Frequently used Delivery Sheaths

- Flexor sheath (COOK) with various modifications
  - Raabe / Anser / Shuttle…
- Others shaths or guiding catheters (Cordis/ Medtronic..)
  - choose compatible internal diameter & length





Techniques

- ✓ **Pre-procedural** : Same as in BPVadequate hydration
- ✓ **Procedure**
  - General anesthesia with endotracheal intubation ± heparinization
  - Otherwise same setting except for stent implantation process
  - Usually pre-dilate RVOT before stenting (make RVOT crossing easier)
  - Stent selection
    - Diameter: 1~2mm larger than PV annulus* or diastolic infundibular diameter#
    - Length: to cover PV to proximal infundibulum*#, or to cover below PV to infundibulum$

      **Castleberry CD. Pediatr Cardiol 2014, #Dohlen G. Heart 2009, $Stumper O. Heart 2013.*
  - Position a long sheath across RVOT → delivery of stent thru LS
  - Checking angio using side arm in the LS during & after stent implantation
- ✓ **Post-procedural**
  - Same as in BPV + usually ICU admission

Outcome / Complications

✓ Efficacy

- SaO2↑, promotes both PA growth → PA index↑
  : typically SaO2 around 70% → around 90%

Dohlen G. Heart 2009, Stumper O. Heart 2013, Castleberry CD. Pediatr Cardiol 2014

- Short~mid-term patency usually sustained until subseq surgery
- Redilation / 2nd stent may be required forlonger-term palliation

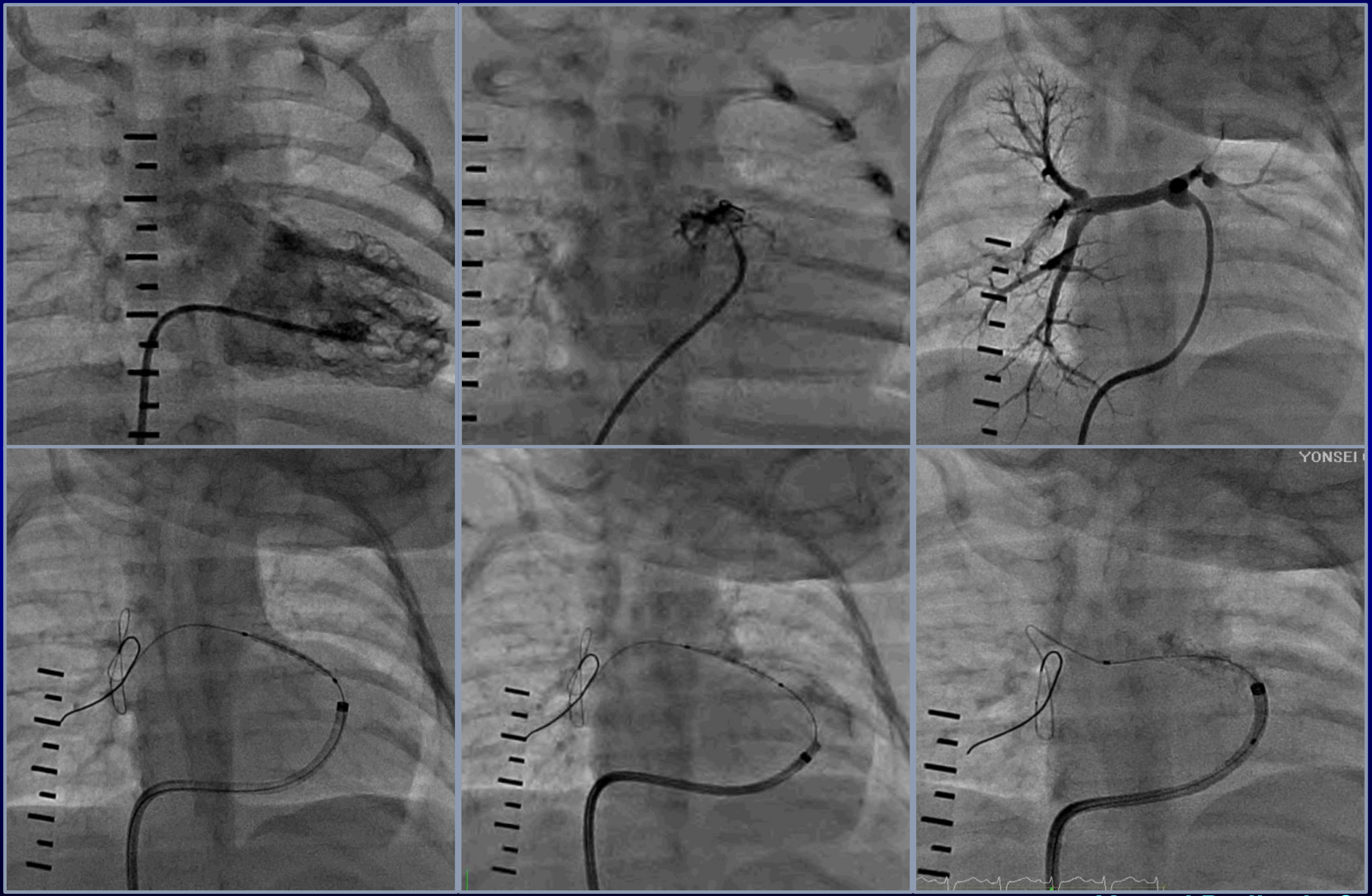
✓ Complications

- Procedure-related mortality : only 1 reported case
- stent migration(& TV damage), emergency surgery (~1.9%)
- Potential Cx : spell/arrhythmia/hypotension during procedure, stent stenosis/thrombosis, stent fracture, vascular/valve damage..
- Other problems
  - sacrifice of PV, complicating subsequent surgery…

Case KBR (F/1mo)

- TOF with nearly atretic RVOT / 2 MAPCAs
- Predilation cordis aviator balloon 5mm

  → *RVOT stent (Core stent 3.5x18mm, dilated to 5mm)*



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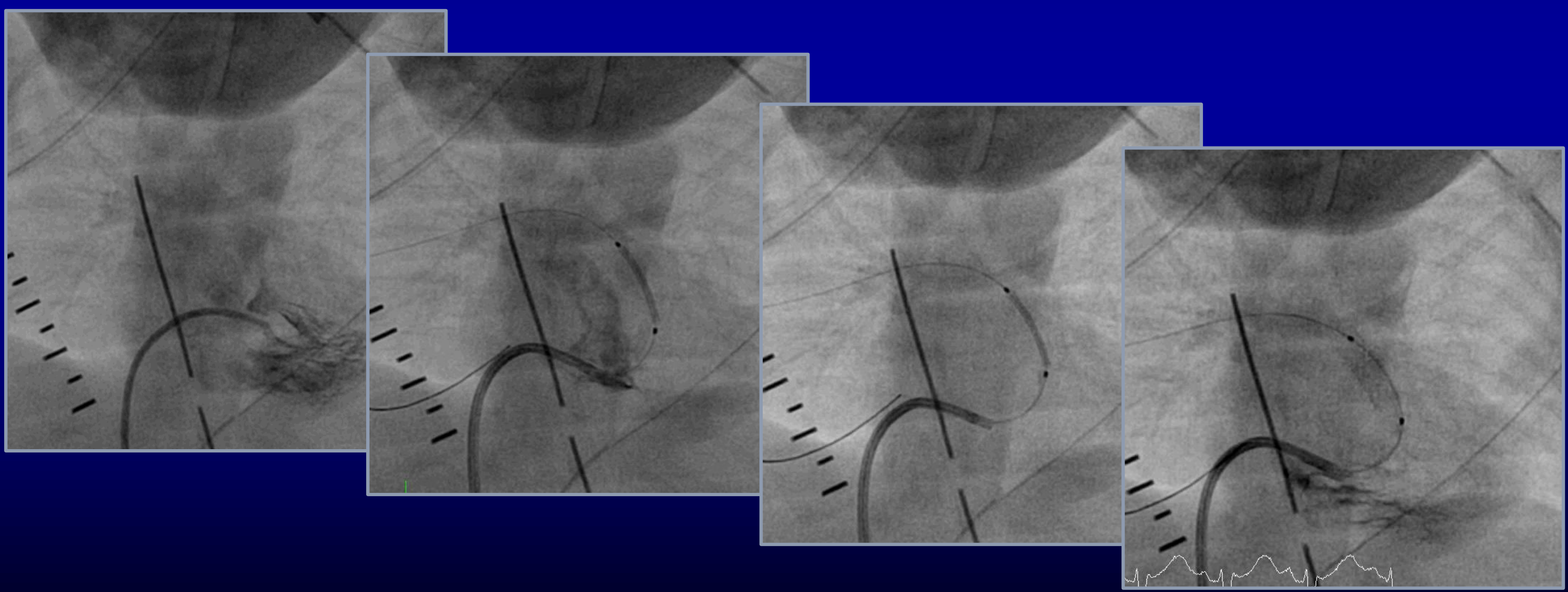
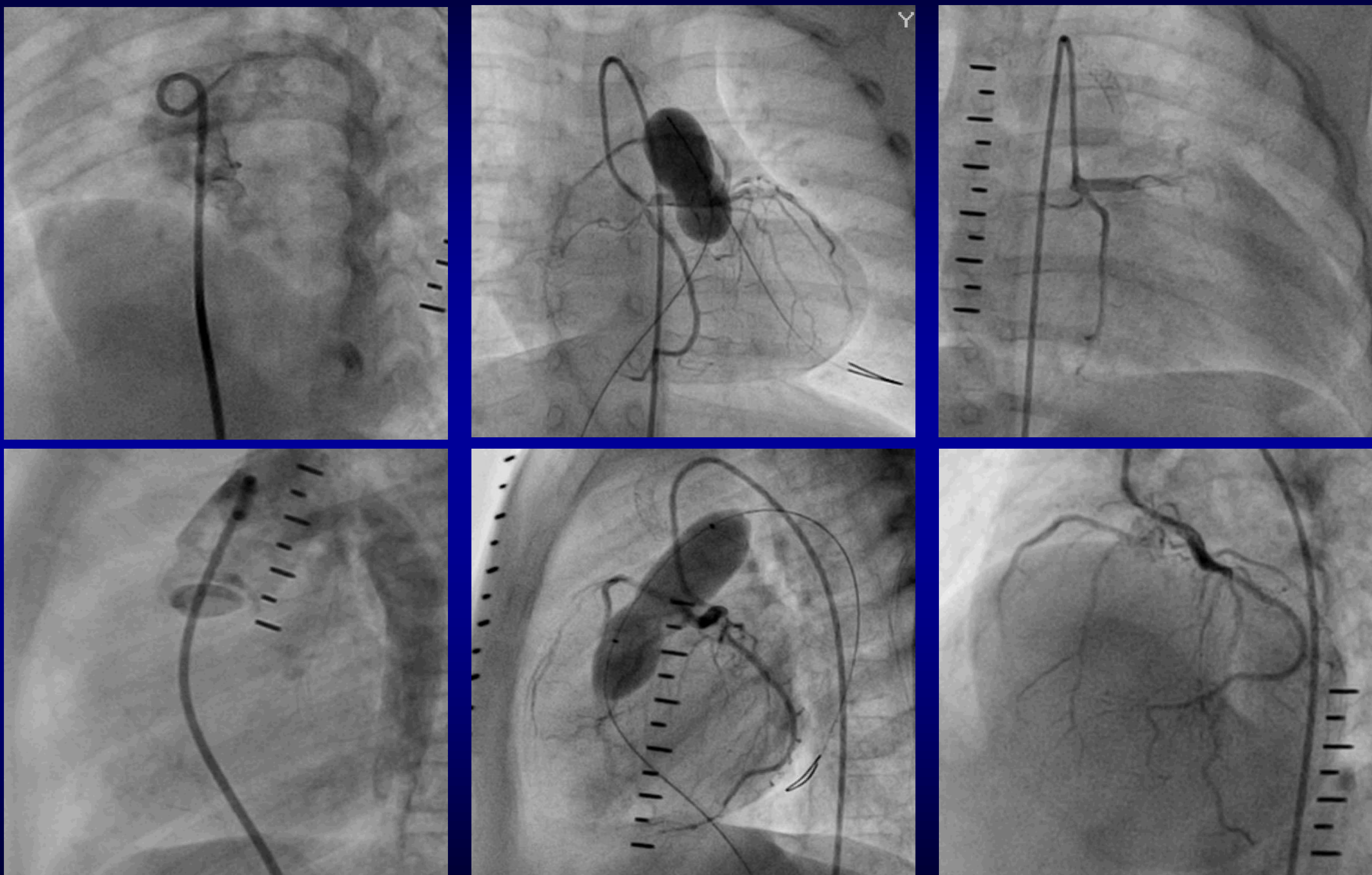
Case KSH (M/1mo)

- repeated episode of severe desaturation since birth
- palliative RVOT relieve at 2wks of age
  → RVOT stent on 26days of age
  (Driver 4.0mm x 15mm, dilated to 6mm)
- total repair at 3mo of age



coronary anomalies



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Cutting Balloon Dilation

Use of Cutting Balloon for Palliative Treatment in Tetralogy of Fallot

Karina M. Carlson, MD, Steven R. Neish, MD, Henri Justino, MD, Glenn T. Leonard, Jr, MD, Charles E. Mullins, MD, and Ronald G. Grifka,* MD

Catheter Cardiovasc Interv 2005;64:507

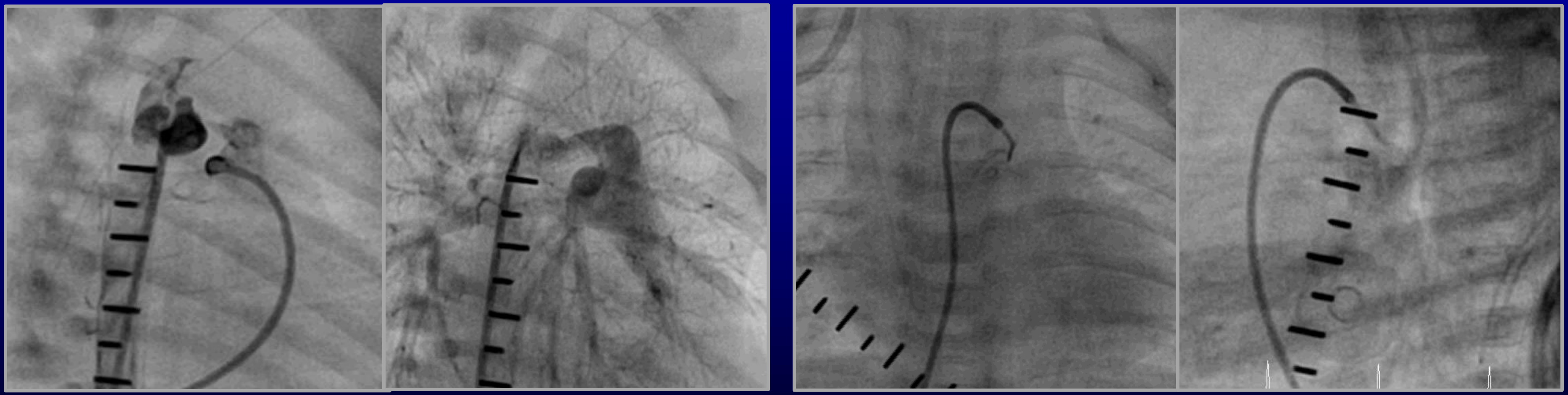
- **4 patients: Critically ill neonate (2.5kg) / 1.5mo infant / 4mo infant (7.5kg) / 20mo child (10.7kg)**
- **Cutting balloon – infund / PV / supravalv portion**
  → standard balloon dilation with 7~8mm balloon
- **Dramatic improvement with favorable course without significant complication**

PDA Stenting

PG dependent pulmonary circulation has been one of the important indication for RVOT stenting
→ PDA stenting may possibly be a reasonable alternative

✓ **PDA stenting is seldom indicated in TOF**

- **Anatomy of PDA in TOF: different from PDA in PA/IVS (short/straight/typical course vs long/tortuous/vertical course)**
- **Presence of easier/safer/verified alternative procedures: balloon dilation / RVOT stenting)**



Conclusion

- ✓ Palliative catheter interventions may be a safe and efficacious alternative to surgical palliation in selected young infants with symptomatic TOF
- ✓ Meticulous approach and decision making:
  - *case selection*
  - *type of intervention*
  - *proper utilization of equipment*
  - *experienced hands*
  - *careful peri-procedural care*
- ✓ Long-term influence on the RV/PV function and/or arrhythmia need to be clarified


Thank you for your attention!

Stenting PABS : *How to ?*

- ✓ **Hemodynamic / angiographic assessment**

  anatomic details are important!

  D/L of stenotic segment, D of adjacent normal vessel

  spatial relationship between side branches..

- ✓ **Pre-dilation**

  to check compliance, balloon effect, stability of balloon on dilation

- ✓ **Select appropriate D/L of stent & balloon**

  should know properties of stents / balloons!

  refer to anatomy of lesion & adjacent normal vessel diameter

- ✓ **Stent delivery**

  usually through a long sheath / lesion-type specific tips helpful

  angiogram to check stent position before balloon inflation

- ✓ **Post-implantation assessment**

Modes of Stent Delivery

- **Through a long sheath**
  - ✓ Stable position - facilitate & secure stent delivery,
    & stabilize stent during deflated balloon removal
  - ✓ Repeated angiography throughout the procedure
    - check stent position before balloon inflation
  - ✓ Easy exchange of catheters / wires & balloons
  - ✓ Useful for post-implant evaluation after stenting.. etc.
- **Over-the-wire without long sheath**

  pre-mounted stent

  (possibly with hand-mounted stent, not recommended)
- **Intraoperative (hybrid)**

Stent Delivery through Long Sheath

✓ **Conventional**

stent delivery through the long sheath after positioning of sheath tip beyond target lesion

✓ **Front-loading technique**

advance stent-balloon-sheath assembly as a unit through the wire which is positioned distal to the lesion

Venczelova Z. CCI 2011

Advantages of Stenting

- **Excellent relief of stenosis**
- **Low restenosis rate**
  - Angioplasty: Imm. success 53-72% / Restenosis 16-35%
  - Stent implantation: Imm. success 95-100% / Restenosis 3-12%
- **Possible staged rehabilitation (serial redilation)**

*** *Disadvantages***

- Technically demanding (learning curve)

- Risk of complications

- Limitations in small infants

Complications of Stenting

Stent malposition / embolization (7.7%)
Jailing of side br. / Restenosis (2%) / Intimal ingrowth(1.8%)
Hemoptysis (1.5%) / Stent fracture / Stent thrombosis
Reperfusion injury - pulmonary edema (1.5%)
Aneurysm formation / Balloon rupture on inflation
Rupture or dissection of vessel / Death (1.5%)

McMahon et al, Cardiol Young 2002

*** Reviews from multicenter registry (US, Europe)**

- ✓ **Complication rate : 19-22% (major 8-10%, death 2.3%)**
- ✓ **Risk factors**

operator experience / patient age, size / hand mounting
pre-existing hemodynamic vulnerability

Holzer RJ. Circ Cardiovasc Interv. 2011;4:287
van Gameren M et al. Eur Heart J 2006;27:2709

Complete Repair of Tetralogy of Fallot in the Neonate

Results in the Modern Era

Jennifer C. Hirsch, MD, Ralph S. Mosca, MD, and Edward L. Bove, MD

From the Section of Cardiac Surgery, Department of Surgery, The University of Michigan School of Medicine, Ann Arbor, Michigan

Annals of Surgery 2000

- **61 symptomatic neonates with TOF (24, pulm atresia)**
- **TAP 49/61 (80%), RV-PA conduit 12/61 (20%)**
- **ICU stay 9.1days / Mechanical ventilation 6.8days**
- **1 hospital death, 4 late death**
- **5yrs survival 93%**
- **1yr/5yr Freedom from reoperation: 89/58%**

Results of elective repair at 6 months or younger in 277 patients with tetralogy of Fallot: A 14-year experience at a single center

Roxanne E. Kirsch, MD,[a] Andrew C. Glatz, MD, MSCE,[b] J. William Gaynor, MD,[c]
Susan C. Nicolson, MD,[a] Thomas L. Spray, MD,[c] Gil Wernovsky, MD,[a,b] and Geoffrey L. Bird, MD[a,b]

JTCS 2014

Early Primary Repair of Tetralogy of Fallot in Neonates and Infants Less Than Four Months of Age

Melanie I. Tamesberger, MD, Evelyn Lechner, MD, Rudolf Mair, MD, Anna Hofer, MD,
Eva Sames-Dolzer, MD, and Gerald Tulzer, MD, PhD

Departments of Pediatric Cardiology, Congenital Heart Surgery, and Anesthesia and Intensive Care Medicine, General Hospital Linz, Children's Heart Centre Linz, Linz, Austria

ATS 2008

Timing of complete repair of non–ductal-dependent tetralogy of Fallot and short-term postoperative outcomes, a multicenter analysis

Matthew B. Steiner, MD,[a] Xinyu Tang, PhD,[b] Jeffrey M. Gossett, MS,[b] Sadia Malik, MD,[a] and Parthak Prodhan, MBBS[a,c]

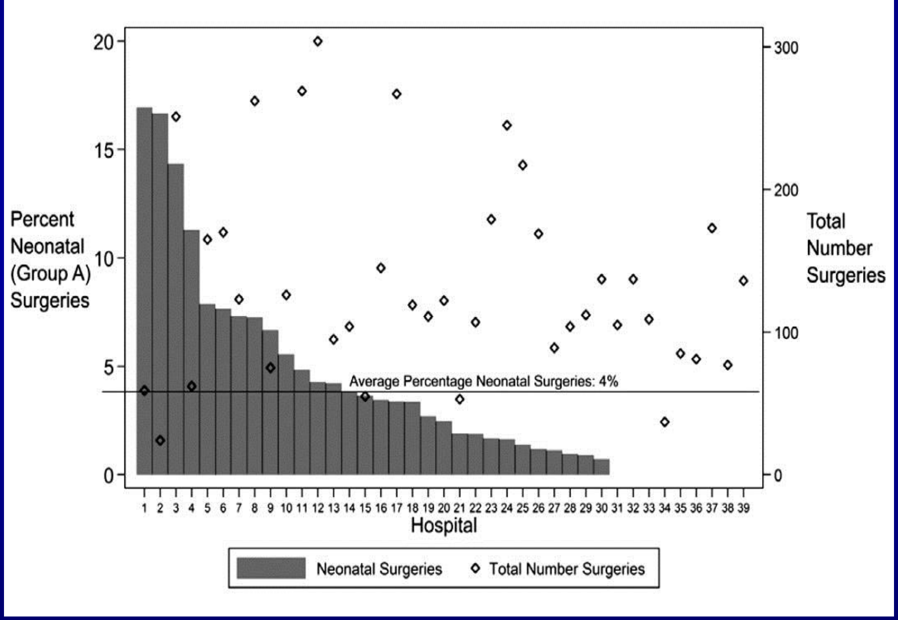
Objective: There is cross-center variability with regard to timing repair of non–ductal-dependent tetralogy of Fallot (TOF). We hypothesized that earlier repair in the neonatal period is associated with increased mortality and morbidity.

Methods: This was a retrospective analysis of the Pediatric Health Information System of tetralogy of Fallot patients undergoing complete repair from 2004 through 2010 between the ages of 1 day to younger than 19 years. Patients with pulmonary valve atresia, those who received prostaglandin during hospital admission, and those who underwent prior shunt palliation were excluded.

Results: A total of 4698 patients met our inclusion criteria, of whom 202 were younger than 30 days old (group A), 861 were 31 to 90 days old (group B), 1796 were 91 to 180 days old (group C), and 1839 were older than 180 days (group D). In-hospital mortality, intensive care unit length of stay, and total hospital length of stay were significantly higher in group A. Patients in group A had a significantly increased postoperative requirement for mechanical ventilation, intravenous blood pressure support, medical diuresis, extracorporeal membrane oxygenation, gastrostomy tube insertion, heart catheterization, and surgical revision. Significant institutional variability was noted for timing of TOF complete repair, with one third of the centers performing 75% of the repairs at younger than 30 days old. The institutional approach to timing TOF complete repair showed no relation to surgical volume.

Conclusions: Across all centers analyzed, primary neonatal elective TOF repair (<30 days of age) is associated with significantly higher postoperative in-hospital morbidity and mortality, although a few centers have shown an ability to use this strategy with excellent survivability. (J Thorac Cardiovasc Surg 2014;147:1299-305)

Group A: 0-30 days (N = 202)	Group B: 31-90 days (N = 861)	Group C: 91-180 days (N = 1796)	Group D: 181 days to 18 years (N = 1839)



Our data confirm our hypothesis, suggesting that, across all centers, complete primary repair of acyanotic TOF in neonates (<30 days old) is associated with increased mortality and morbidity. We found a significantly higher overall in-hospital postoperative mortality rate (6.4%) for elective neonatal complete repairs. In addition, total billed hospital charges, the total hospital LOS, ICU LOS, time until extubation, postoperative requirement for revision, medications, and secondary procedures all were increased significantly for elective neonatal complete repairs. There

smaller percentage of their surgical distribution. It was notable that 3 of the 4 centers showing a particular propensity to perform neonatal repairs did not experience any group A mortality whatsoever, indicating that it is possible to achieve excellent survivability when a center has committed to this strategy.[8] The by-center analysis for morbidity, however, still mirrored that seen across the entire cohort, with total hospital LOS, ICU LOS, time on mechanical ventilation, medication requirement, and postoperative procedural complication rates all increased with earlier repairs, even at centers achieving low mortality after neonatal repair.