Ductal stenting: Who, When, How and What thereafter?

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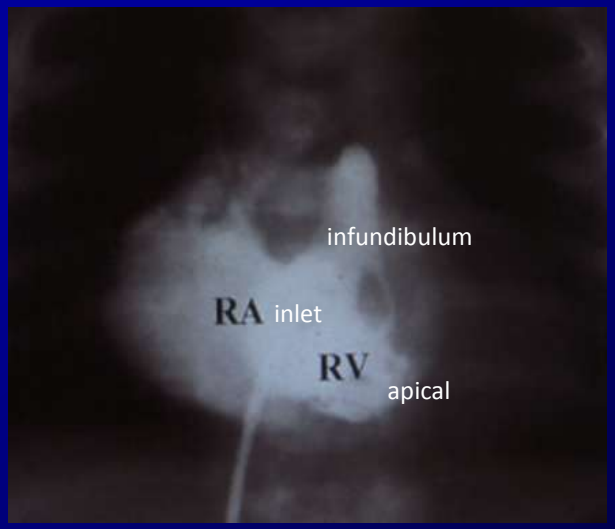
WHO …

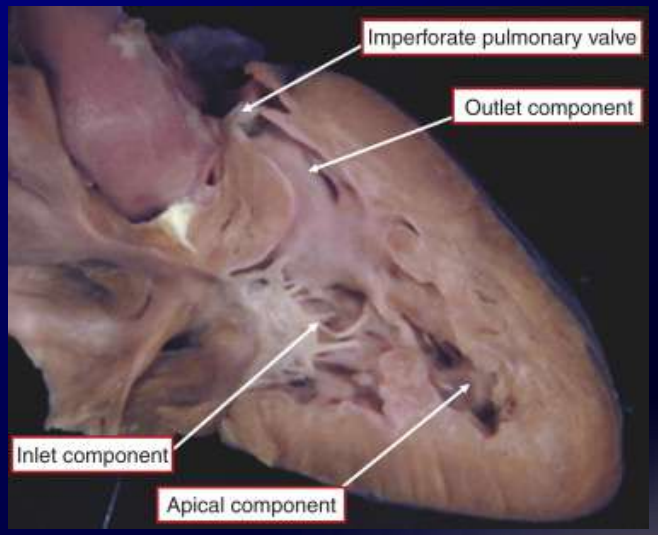
- **PA-IVS : at the time valvotomy & balloon dilatation as additional source of pulmonary blood flow (inadequate RV size, RVH, ↓↓ compliance)**
  (Also severe Ebstein's anomaly with "functional" atresia)

- **(PA-VSD) Tetralogy of Fallot – Pulm atresia: alternative palliation to neonatal BT shunt (also TGA-VSD-PA, ccTGA-VSD, single ventricle with PA)**

- TGA with involuted LV – PDA stent as alternative to BT shunt and PA band for LV-retraining

…A tale of 2 pulmonary atresias





PA-IVS with good anatomy
(tripartite RV, membranous atresia)

- Valvotomy + balloon dilation – may be the only procedure required for the medium term

  (Long term – severe tricuspid or pulmonary regurgitation may require re-interventions)

Anderson et al. Paediatric Cardiology, 3rd ed. 2010

Humpl T et al. Circulation 2003; 108: 826 – 832

27/30 pts – Successful RF-valvotomy

Tricuspid valve z-score -1.33

RV length z-score -5.93

→

14/27 – Unplanned modified BT shunt
2-24 days post RF-valvotomy

2/27 – Unplanned RVOT reconstruction
(<48 hours post)

Agnoletti G et al. J Am Coll Cardiol 2003; 41: 1399 - 403

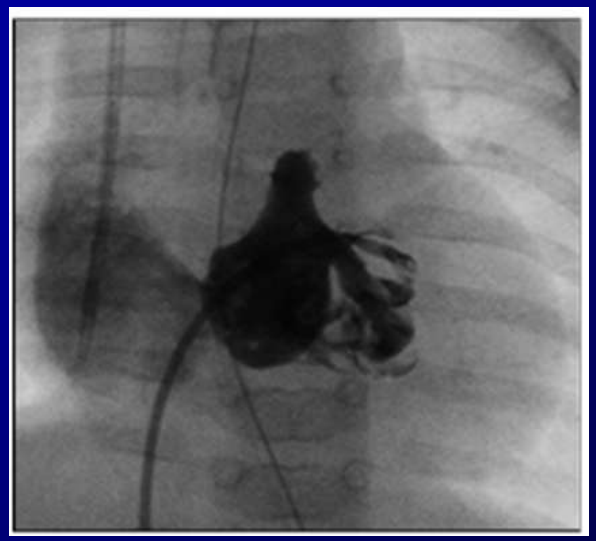
33/39 – Successful RF valvotomy
(mean tricuspid valve -0.5)

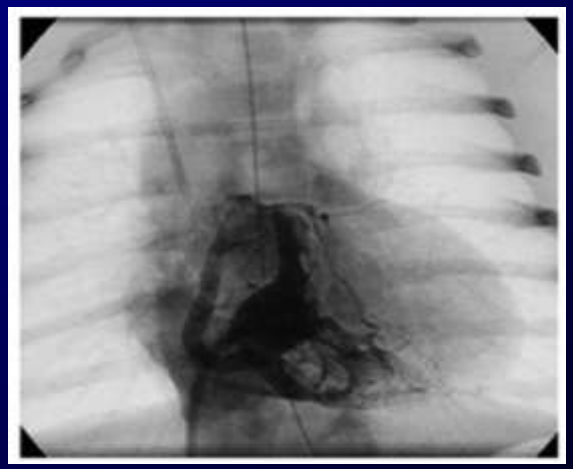
→

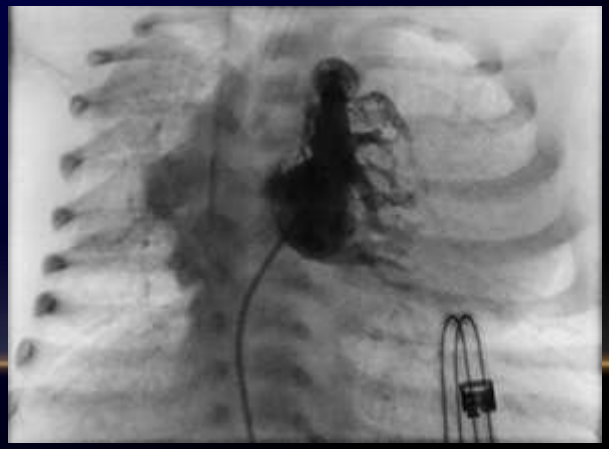
21/33 – Unplanned neonatal surgery (all required continued PG infusion)
12 – BT shunt
8 – BT shunt + RVOT patch
1 – RVOT patch

PA-IVS – Bipartite RV, "non-apex forming"

Common anatomic feature at presentation





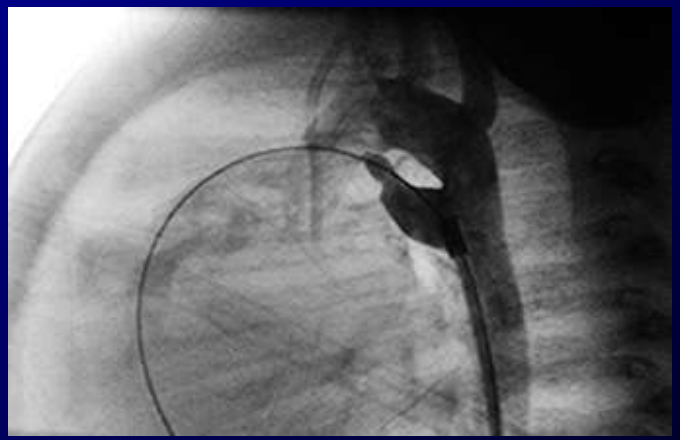


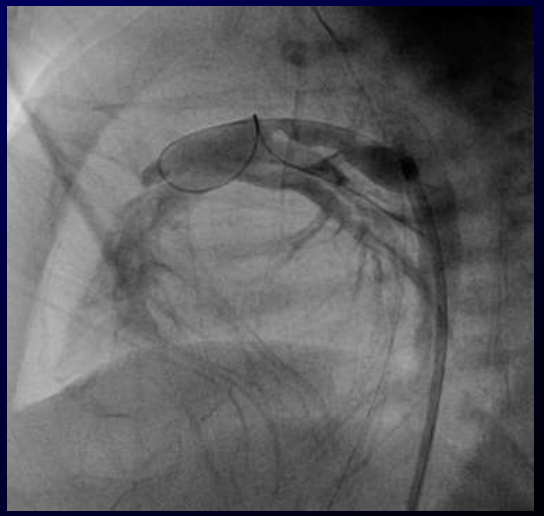
- Small RV cavity
  Bipartite RV – apical part nearly obliterated by muscles (short RV length)
- Muscle-bound RV, poor compliance

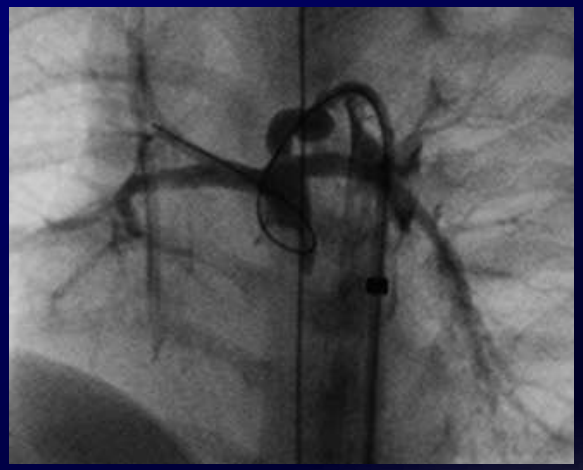
  ↓↓ filling capacity – inadequate as pulmonary ventricle
  Requires additional source of pulmonary blood flow

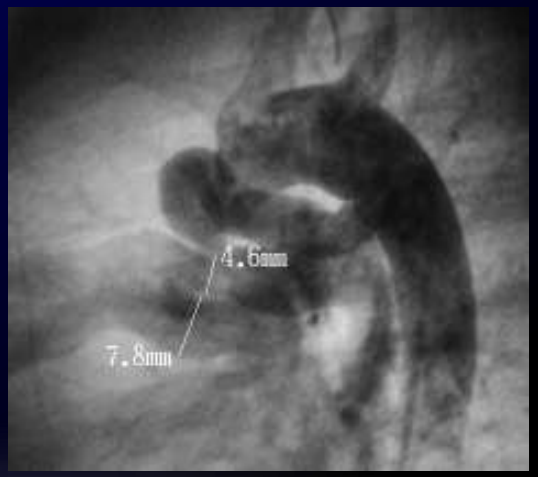
Concomittant PDA stent and RF valvotomy avoids unplanned BT shunt/PDA stent due to continued PGE1 dependency

The PDA in PA-IVS

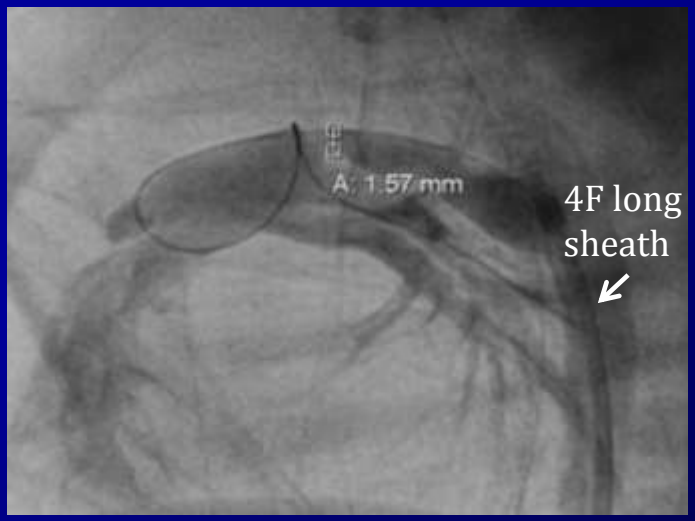








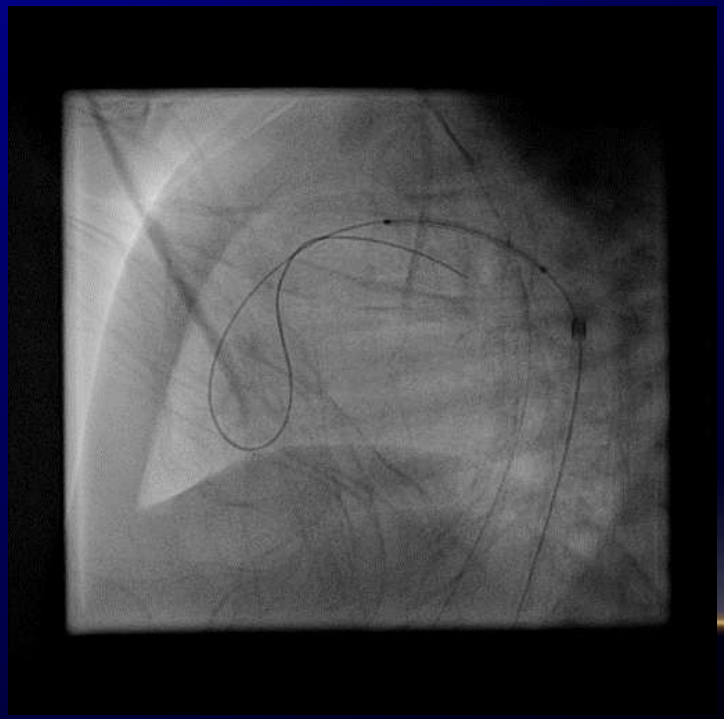
Arises from distal arch/proximal descending aorta, long but not overly tortuous, insert onto dome of main pulmonary artery instead of onto LPA (less problem with LPA stenosis)

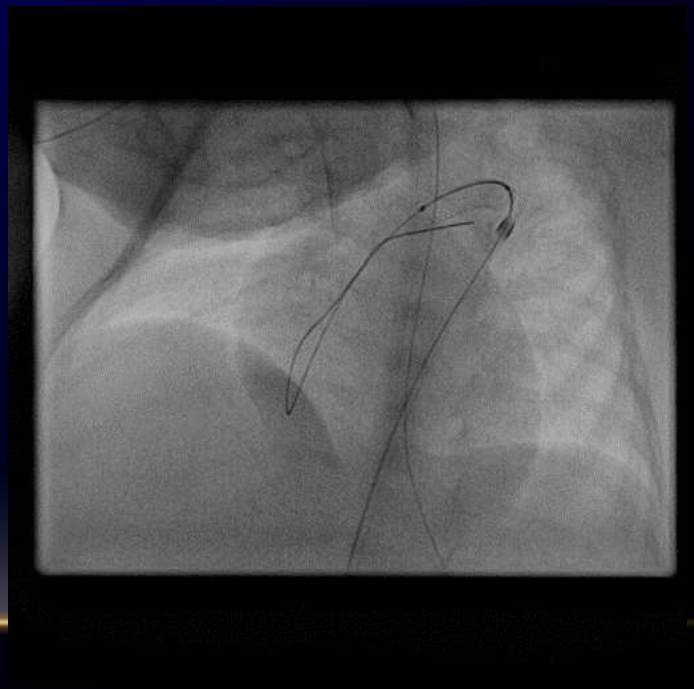


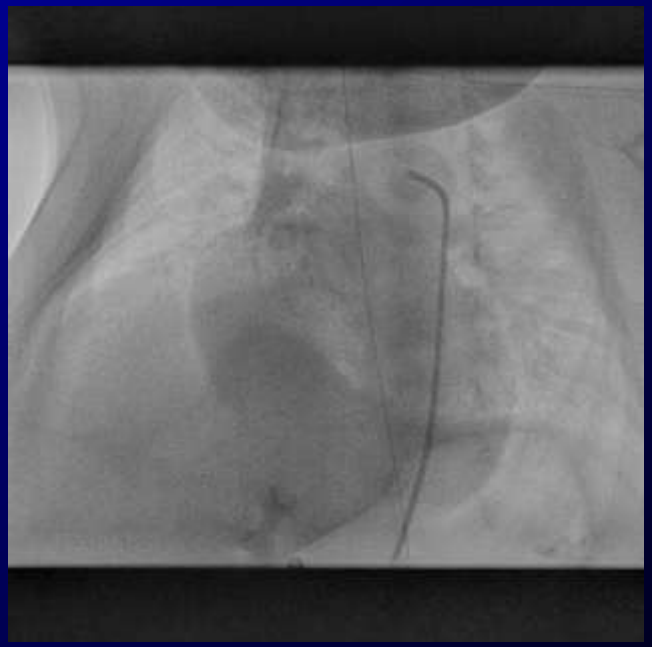
Technique

PA-IVS, bipartite RV
F, 2 weeks old, 3.1 kg

- Valvotomy and balloon dilatation
- PDA stenting, femoral artery route
- Use of 4F Mullins sheath
- 4.0 mm diameter stent for ≥ 3.0 kg infants



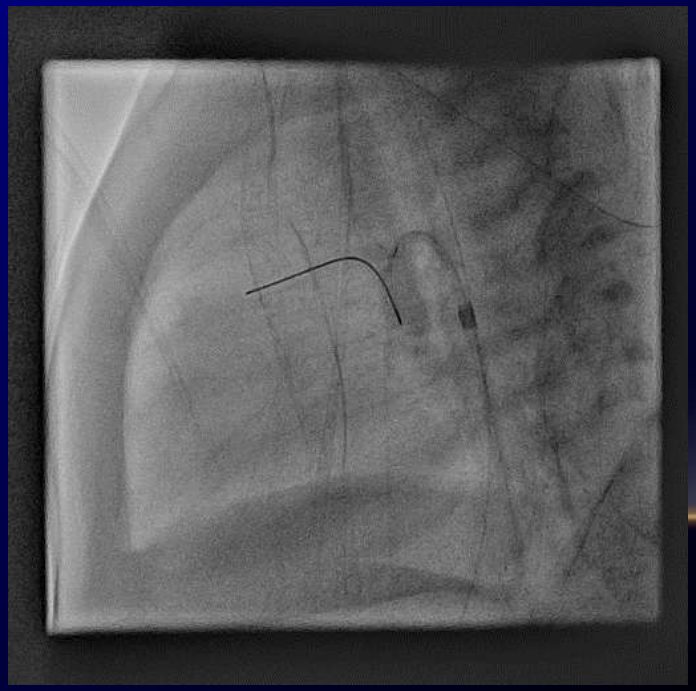


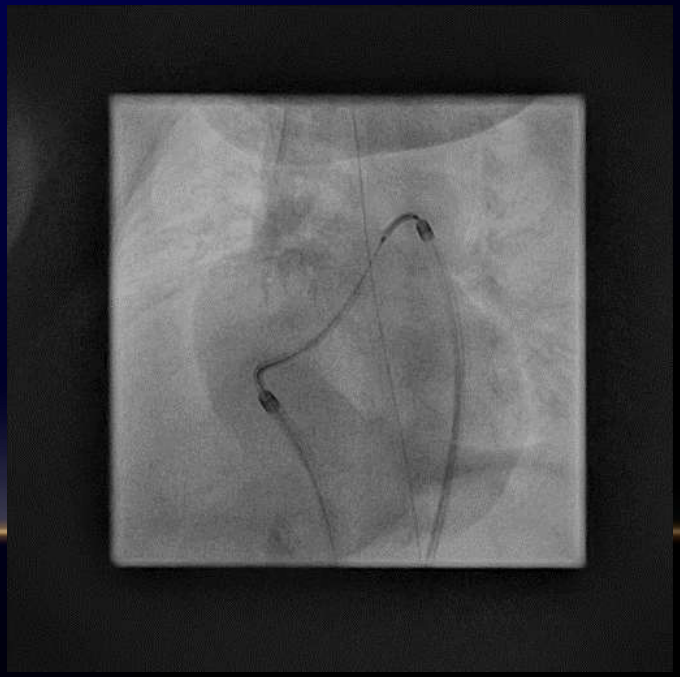


PA-IVS, bipartite RV

RV-LCA communication (not RV dependent coronary circulation), valvotomy + balloon dilation

Long, tortuous PDA

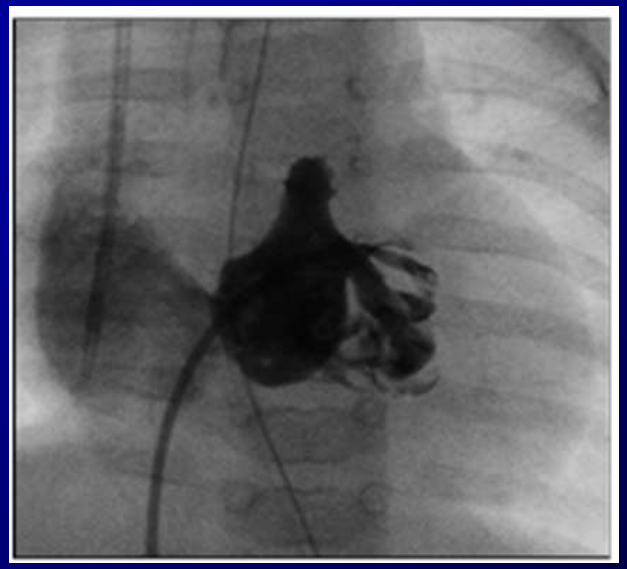




Cut pigtail to engage PDA orifice for floppy wire to cross PDA

Snare assistance for wire stability and stenting

PA-IVS with borderline RV (often bipartite with good RVOT, apical nearly obliterated by muscles)

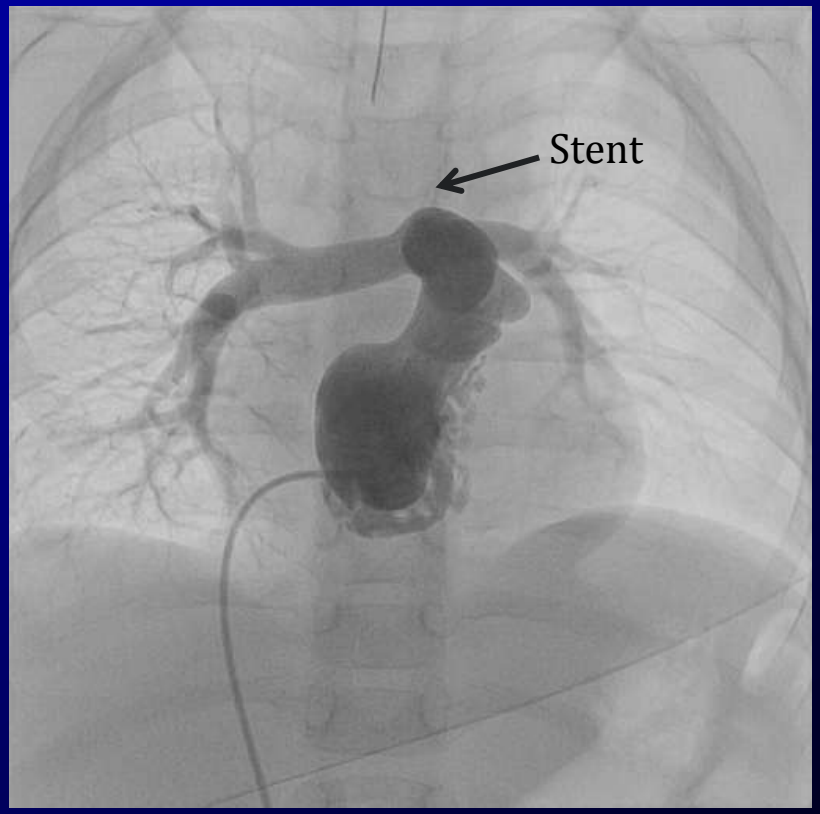


- Transcatheter valvotomy and balloon dilatation : Often abolishes obstruction to RV outflow but cyanosis may remain severe as RV is inadequate to handle total preload
  R → L shunt at PFO
- Concomittant PDA stent as additional source of pulmonary blood flow

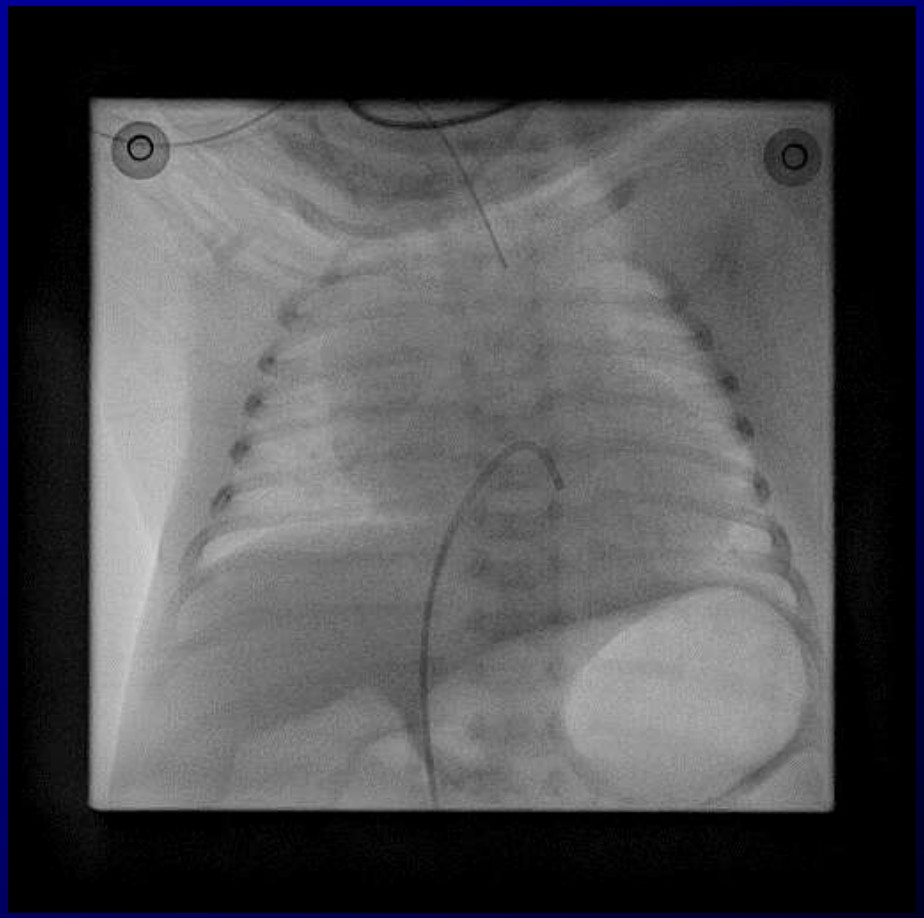
1 – 3 years

- ✓ Sufficient RV growth – normal RV pressure - ↓↓ muscle hypertrophy, ↑↑ RV volume (2 ventricle circulation), minimal R → L shunt (may need device closure of ASD)
- ✓ Insufficient RV growth – inability to handle total cardiac output, still significant R → L shunt, Glenn shunt to off-load RV (1 ½ ventricle circulation)

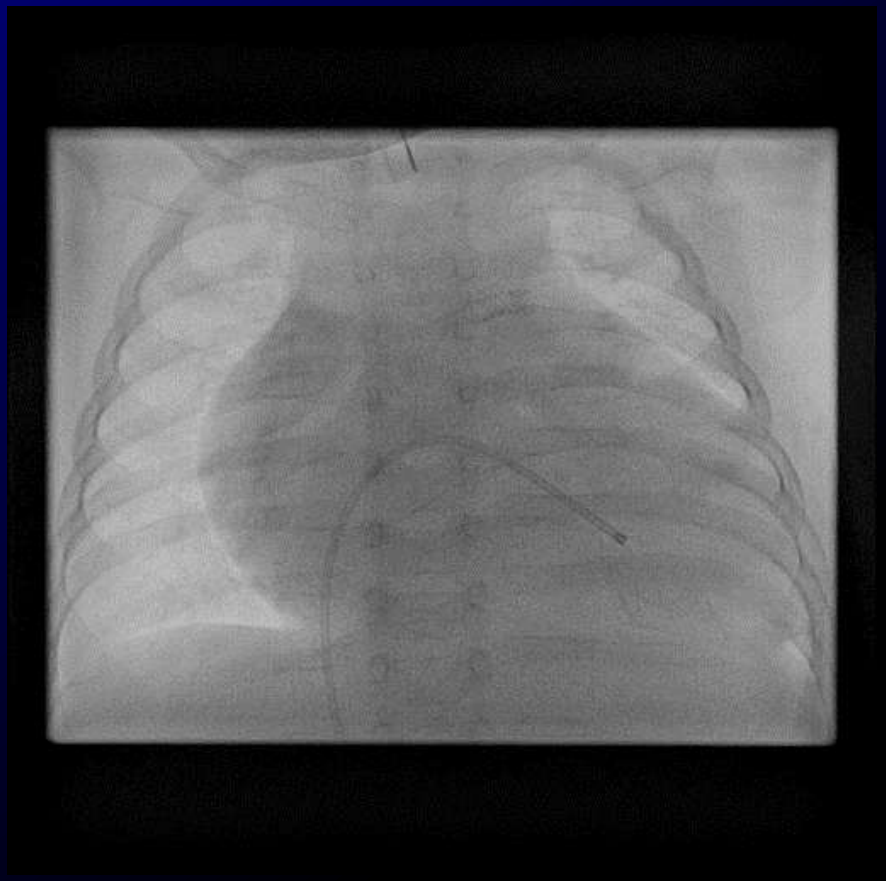
RV fails to grow (non-apex forming)



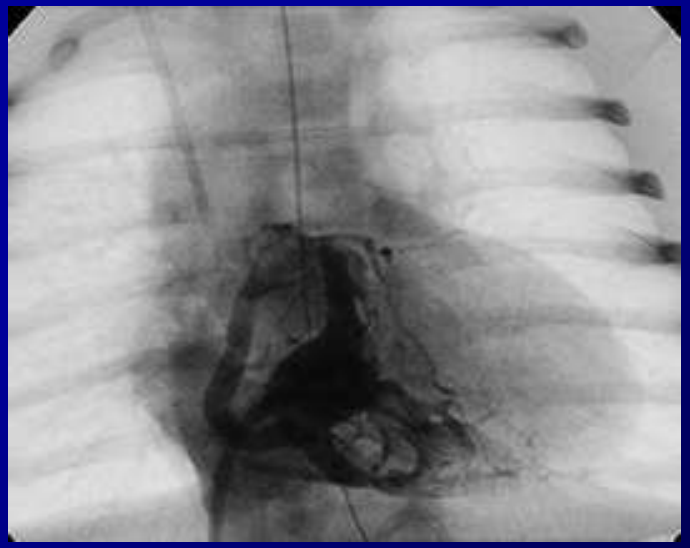
2 years post valvotomy + PDA stenting
RV apex remains under-developed, no RVOT obstruction
PDA stent blocked
Moderate cyanosis
→ Glenn shunt (1 ½ ventricle circulation)

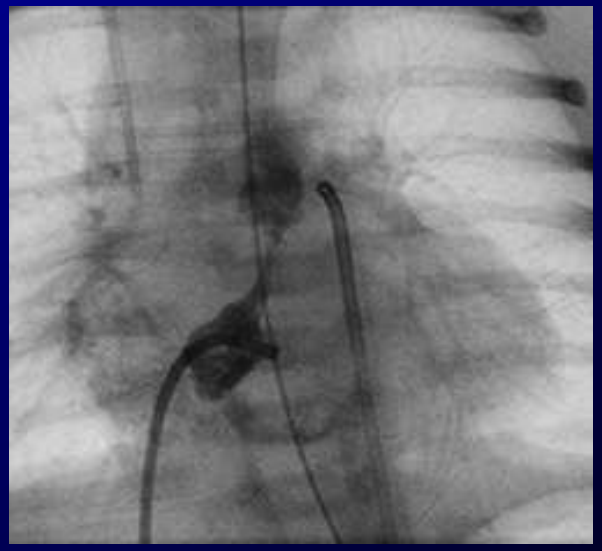


At valvotomy (2 weeks old)

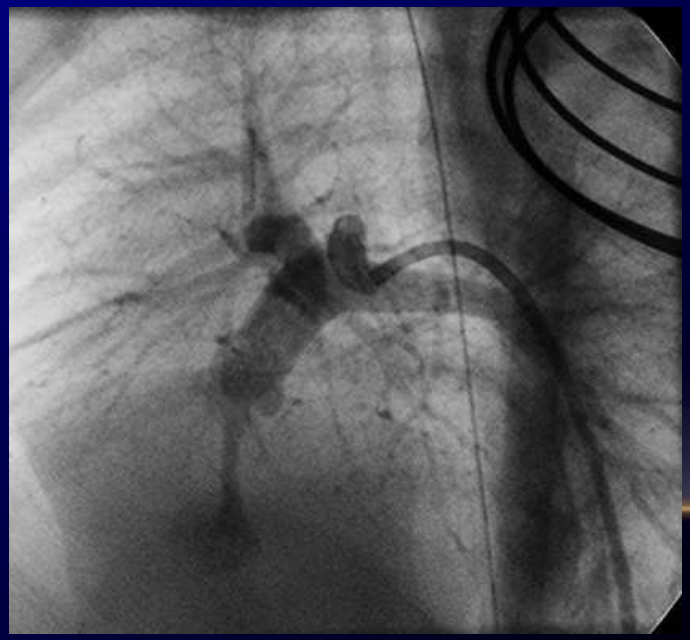


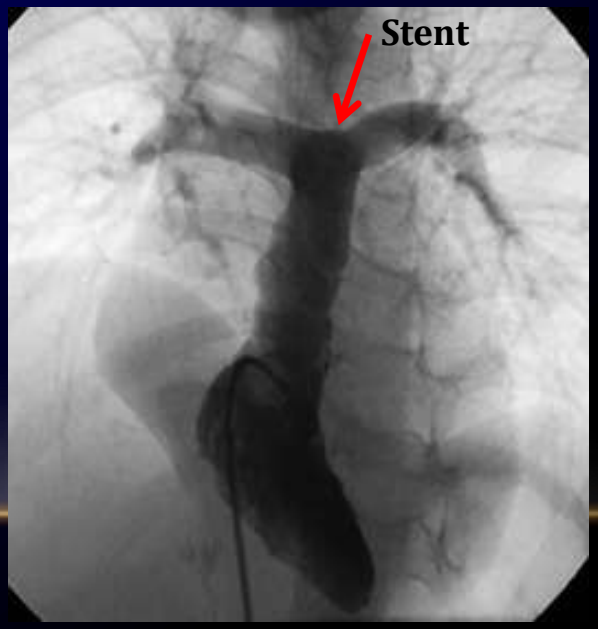
6 months post valvotomy
+ PDA stent





PA-IVS, bipartite RV
Major RV-RCA communication, minor communication to LCA (non RV dependent coronary circulation)
RF valvotomy + PDA stent





4 years post
Self-limiting PDA flow, well developed RV, no RV-coronary communications
Well developed RV (RVOT & apex)

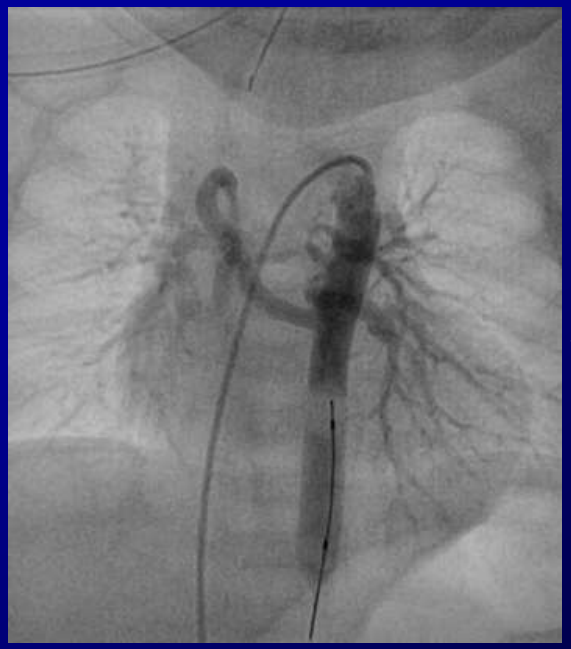
PDA stent – Who …

- **PA-IVS – PDA stent to augment pulmonary blood flow following valvotomy balloon dilation**

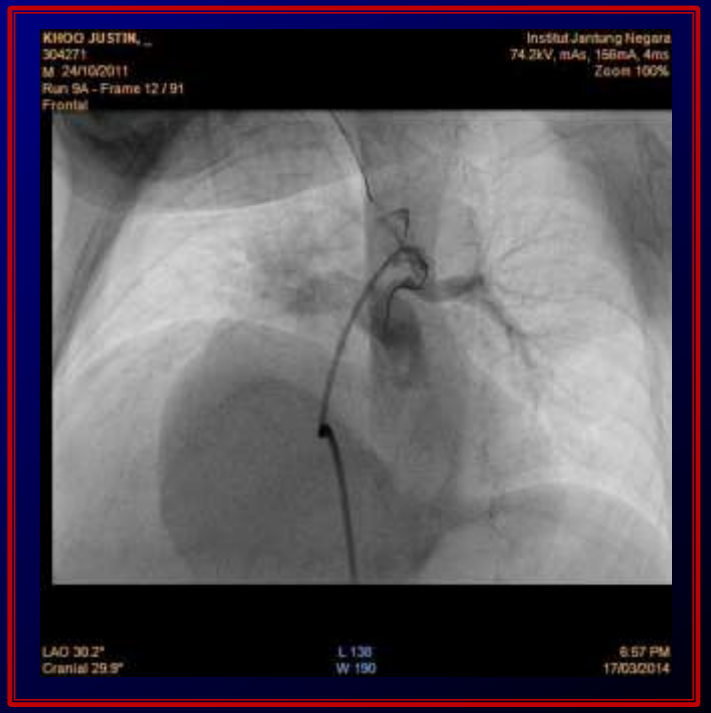
- **TOF-PA – First stage palliation – PDA stent as sole source of pulmonary blood flow**
  (also TGA-VSD-PA, ccTGA-VSD-PA, single ventricle-PA including PA-IVS)

- TGA with involuted LV
  LV re-training : PDA stent as alternative to PA band + BT shunt

TOF-PA

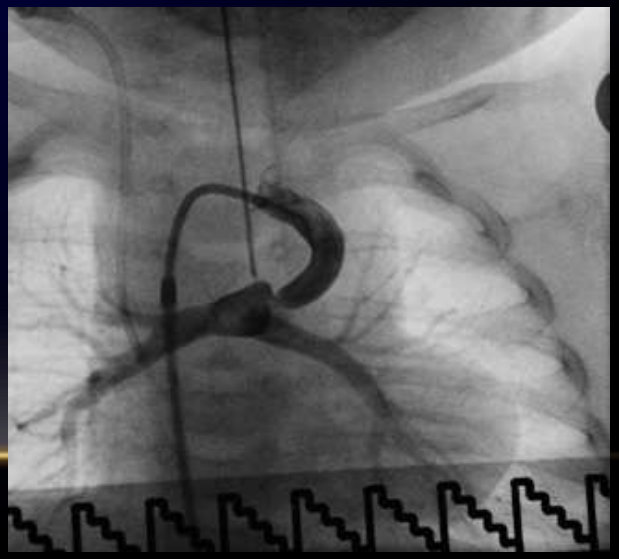


Multiple MAPCAs
Very small native PAs
No PDA

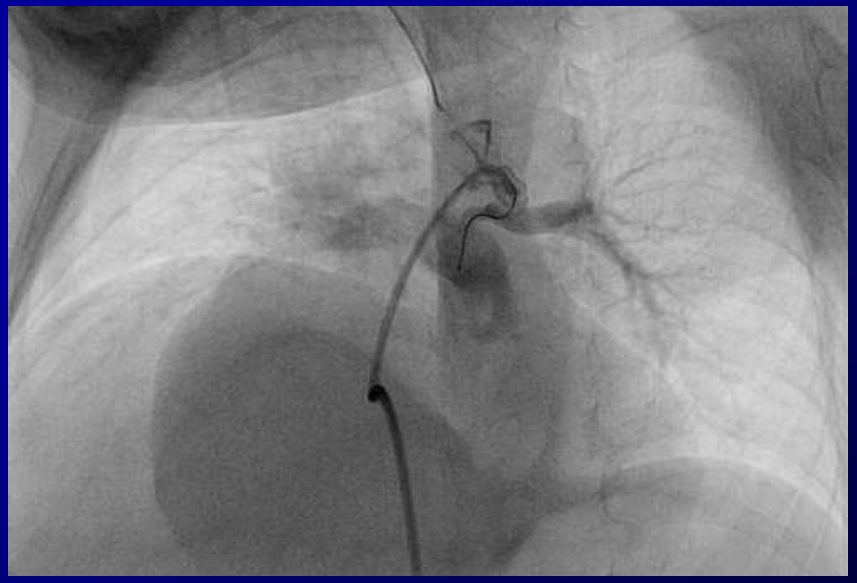


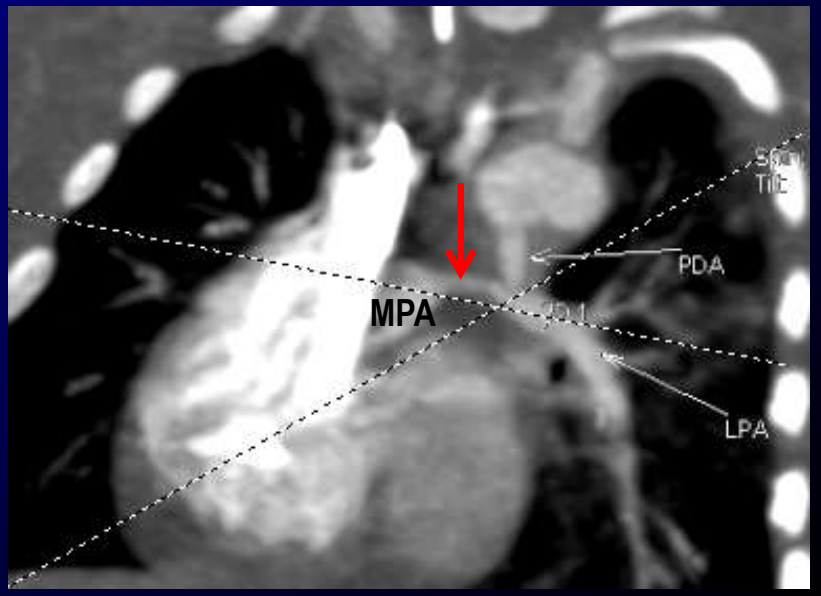
PDA from underside of aortic arch onto LPA

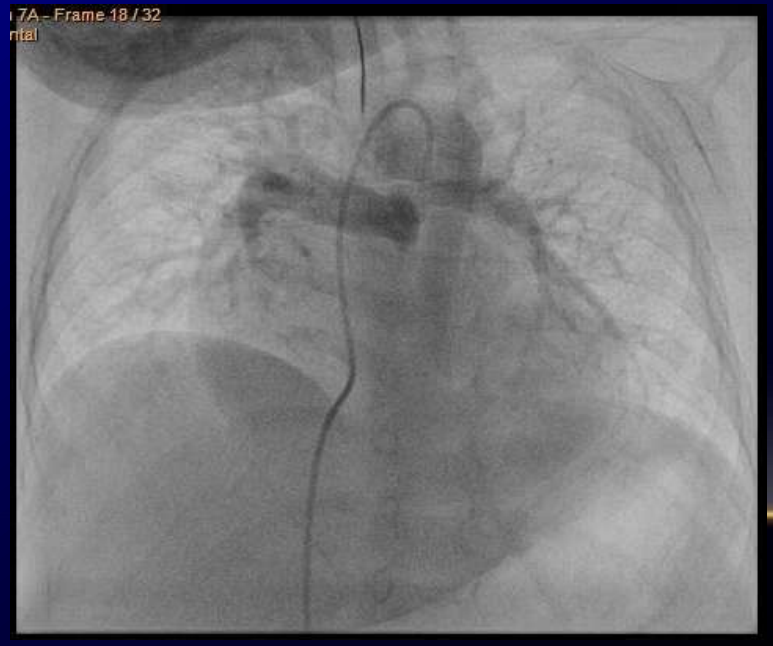
Bilateral PDAs



PDA from base of left subclavian artery







PDA arising from underside of aortic arch, inserting onto proximal LPA, vertical course

The ductus arteriosus and stenoses of the pulmonary arteries in pulmonary atresia

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Elzenga NJ, Gittenberger-de Groot AC. The ductus arteriosus and stenoses of the pulmonary arteries in pulmonary atresia. Int J Cardiol 1986;11:195–208.

J THORAC CARDIOVASC SURG 1990;100:416-24

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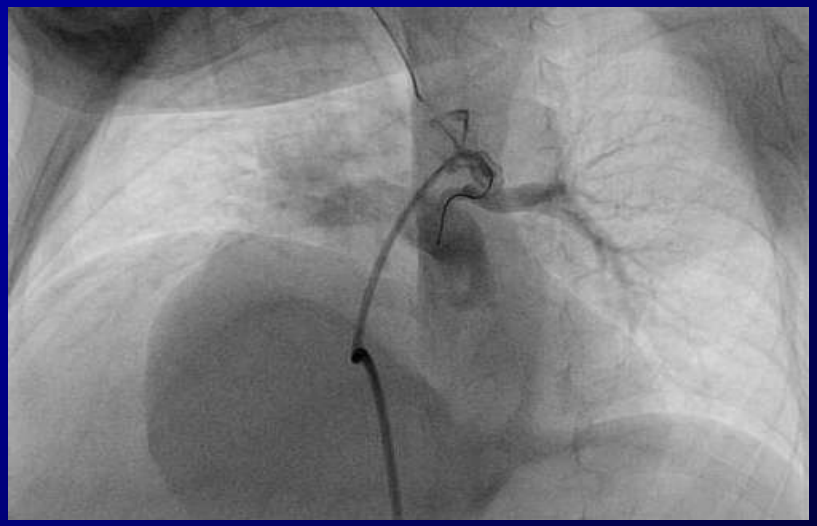
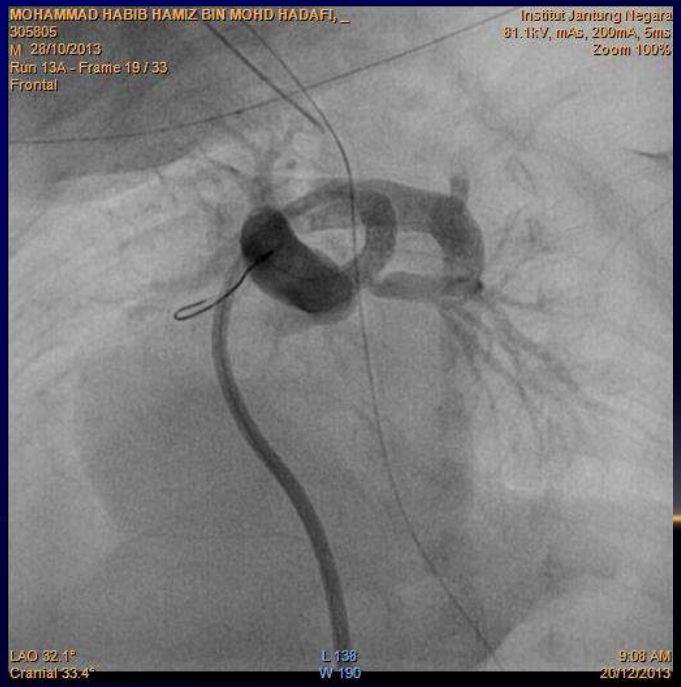
Juxtaductal pulmonary artery coarctation

An underestimated cause of branch pulmonary artery stenosis in patients with pulmonary atresia or stenosis and a ventricular septal defect

An angiographic and clinical study was performed to establish the prevalence of juxtaductal pulmonary artery coarctations in patients with pulmonary atresia or stenosis and a ventricular septal defect or a complex intracardiac defect. The present study is an adjunct to a previously reported portmortem study, in which the incidence of these pulmonary artery coarctations was found to be unexpectedly high. Pulmonary artery coarctations were identified angiographically in 10 of the 15 patients with pulmonary atresia. One additional patient had a bilateral ductus arteriosus and confluent pulmonary arteries, but did not have a pulmonary artery coarctation. Pulmonary artery coarctations were much less prevalent in the cases with pulmonary stenosis (5/50). However, these pulmonary artery coarctations appeared identical to those of the cases with pulmonary atresia. Fourteen pulmonary artery coarctations were located in the pulmonary artery at the side of the ductus arteriosus; this was left

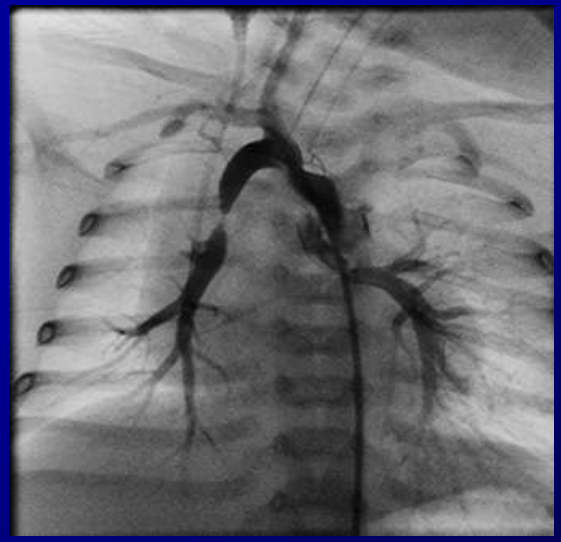
PDA stent in ductus-related pulmonary coarctation





✗ ▪ Jailing of LPA, worsening and accelerated LPA stenosis in response to stent implantation?

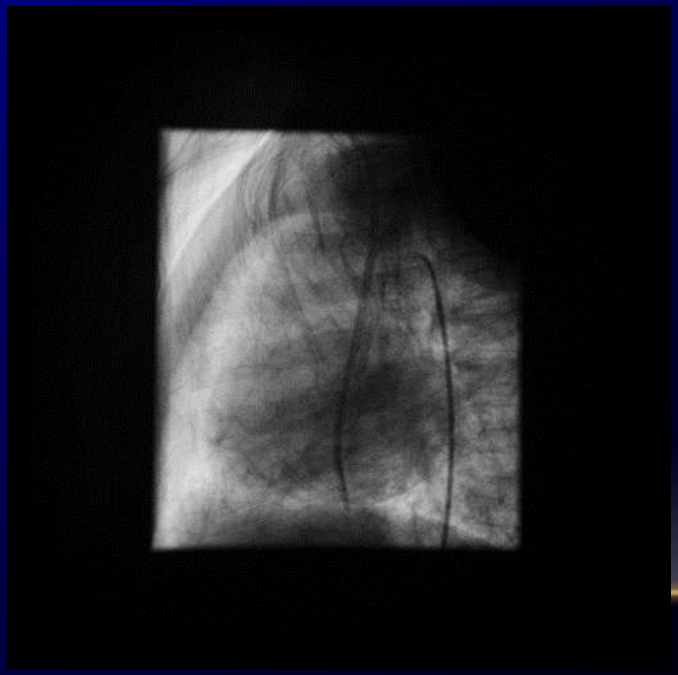
✓ ▪ PDA stent directed towards MPA may limit impact of ductal constriction, prevent total disconnection of LPA

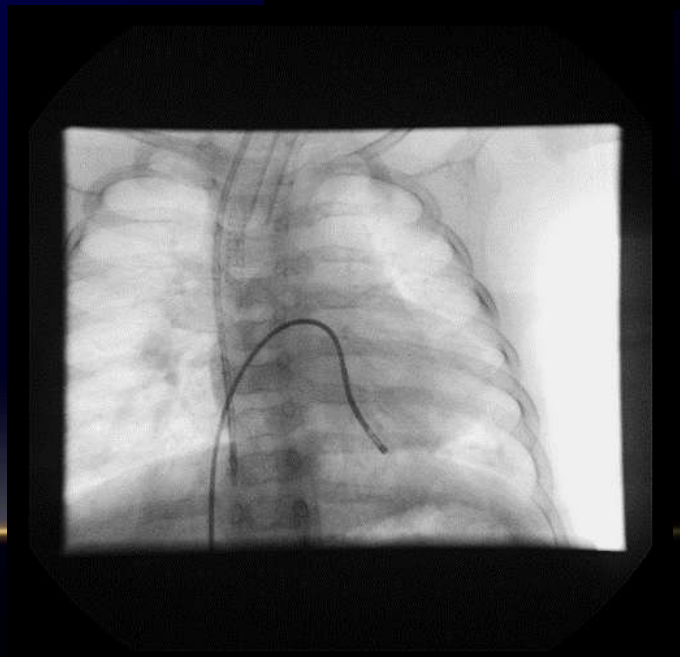


TOF-PA, bilateral PDA

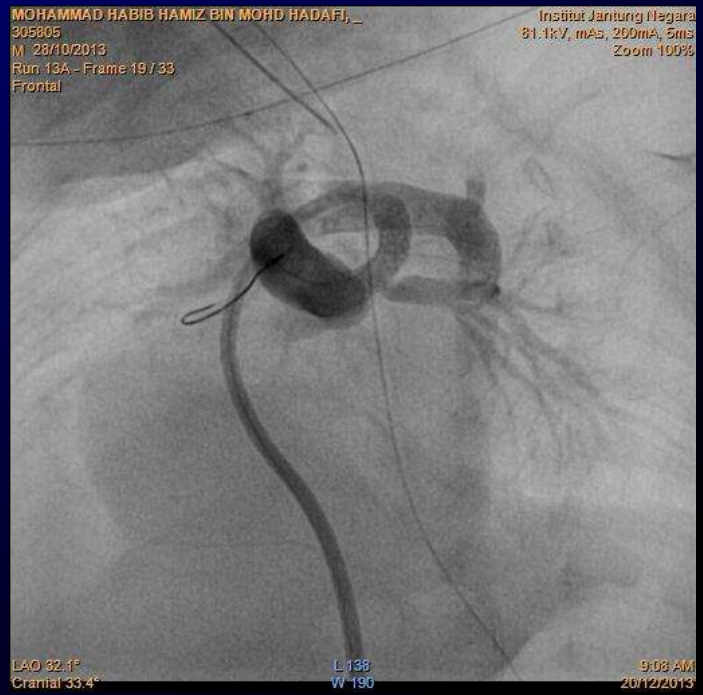
Right PDA stent at 1 month
Left PDA to LPA – large flow

5 months
Left PDA closed

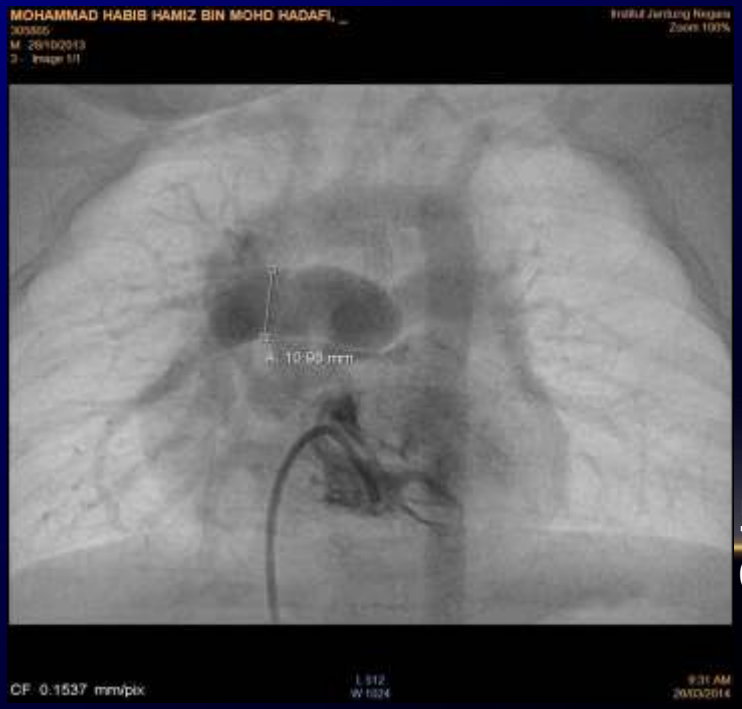




TOF-PA, ductus related LPA stenosis

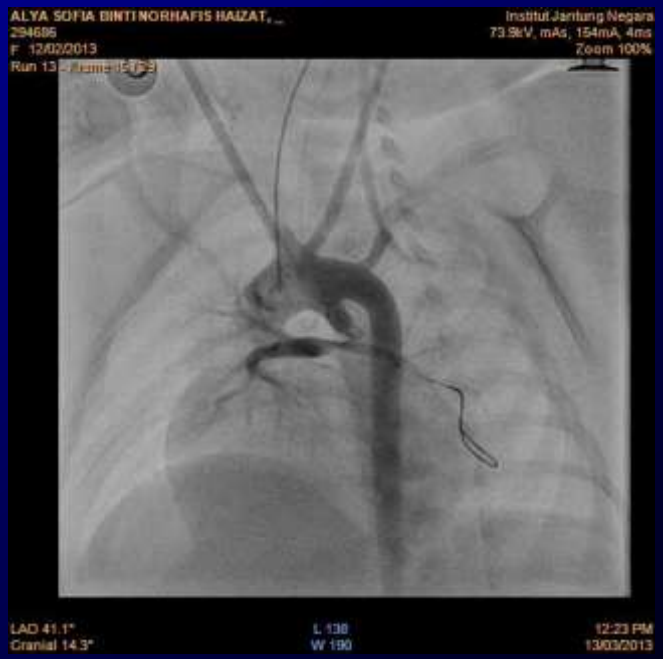


4 weeks of life – PDA stent

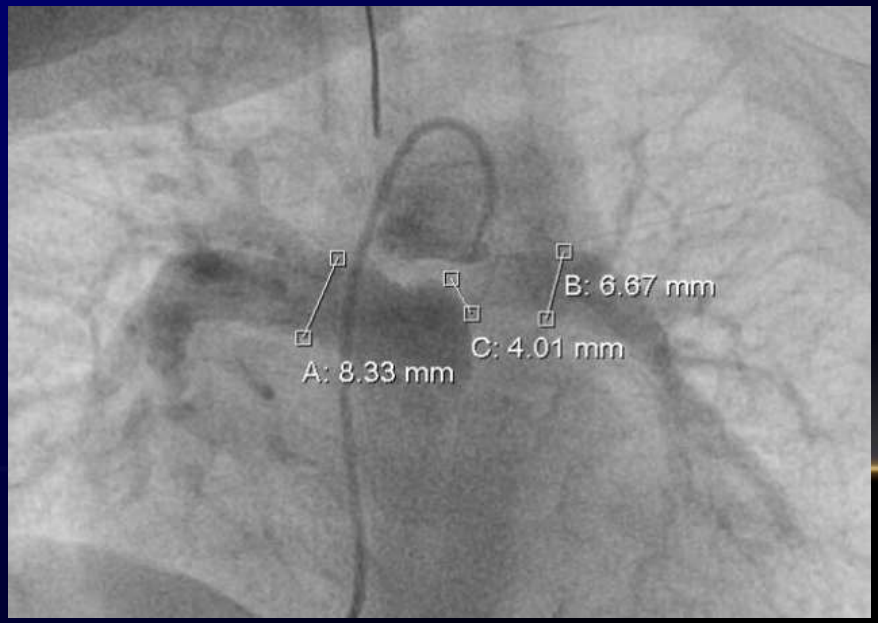


5 months post

Good growth of LPA despite jailing by stent

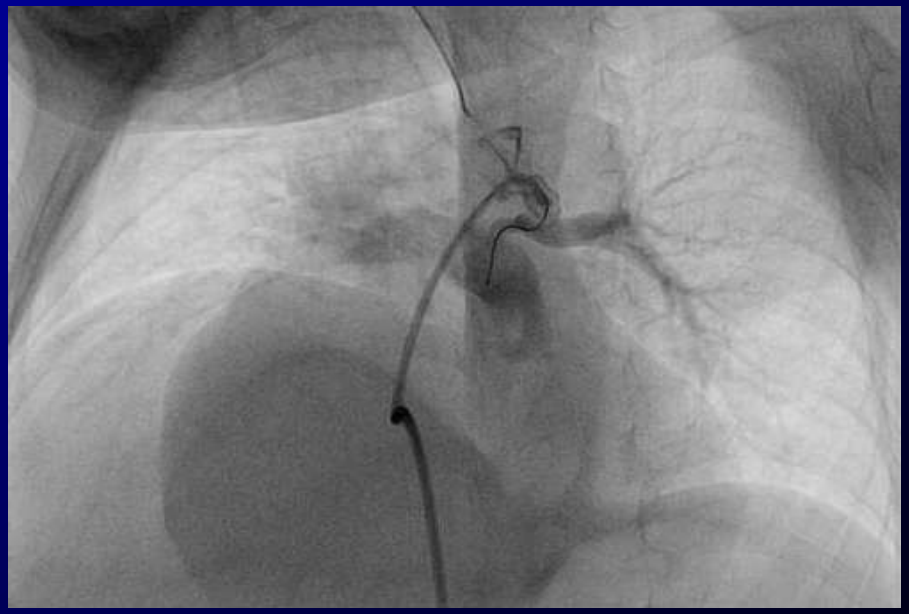


Stenting via right carotid artery cut-down at 3 weeks of life



9 months post PDA stenting
Good growth of LPA

PDA stenting in ductus-related LPA stenosis



MSCT-1
Confirm diagnosis & recruitment

→ Angio & PDA stenting

→ MSCT-2
6-12 weeks

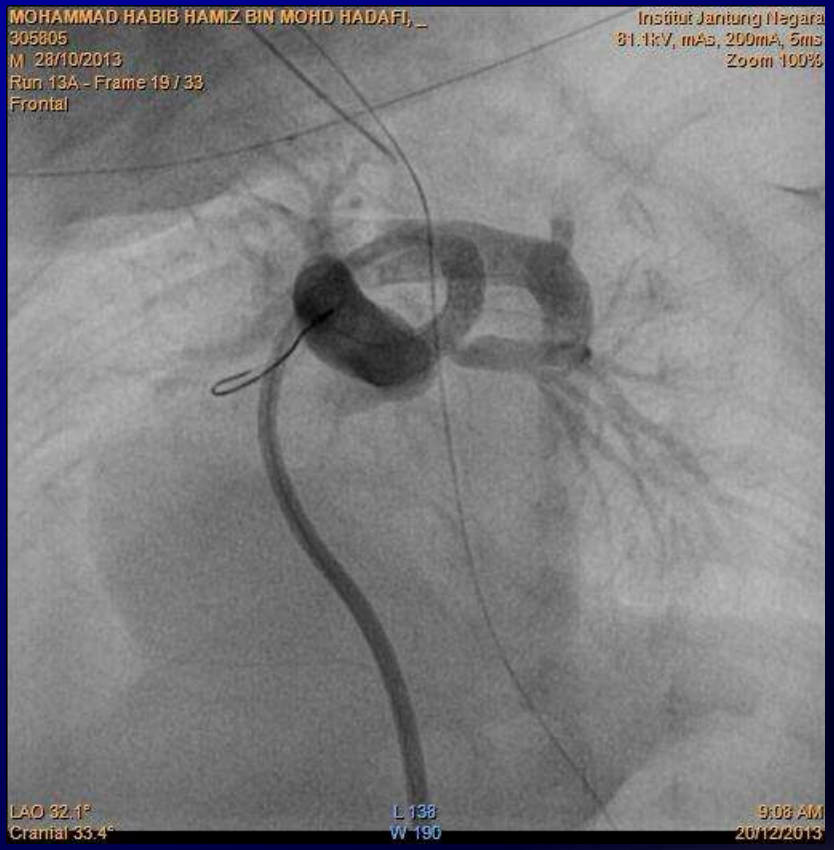
→ Surgical repair
6-9 months

→ MSCT-3
End point

Severe LPA stenosis, small LPA

→ Rescue left BT shunt (end point)

PDA stenting in TOF-PA, ductus related LPA stenosis … How

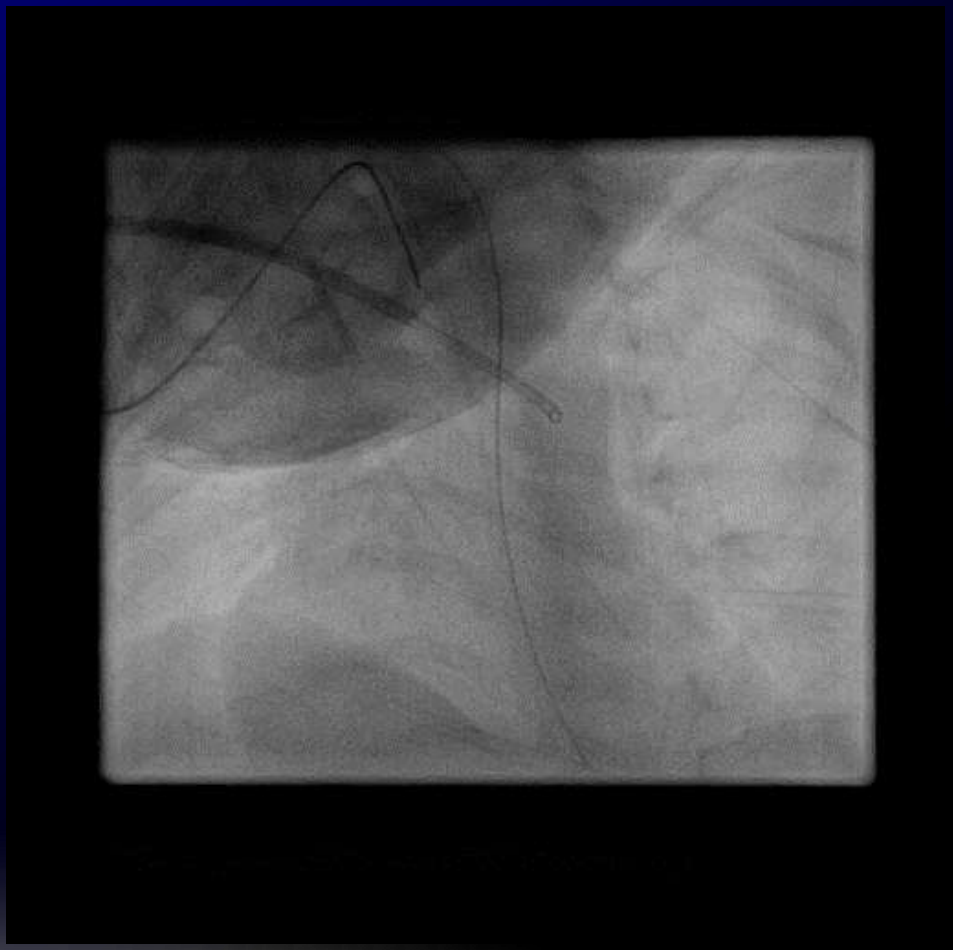


Transvenous, transVSD approach with JR guide catheter

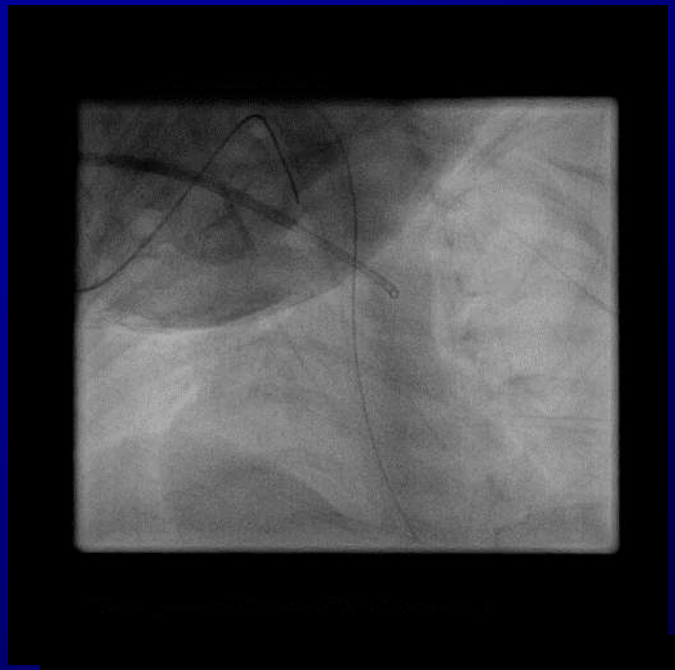
Proximal origin of PDA, from underside of aortic arch

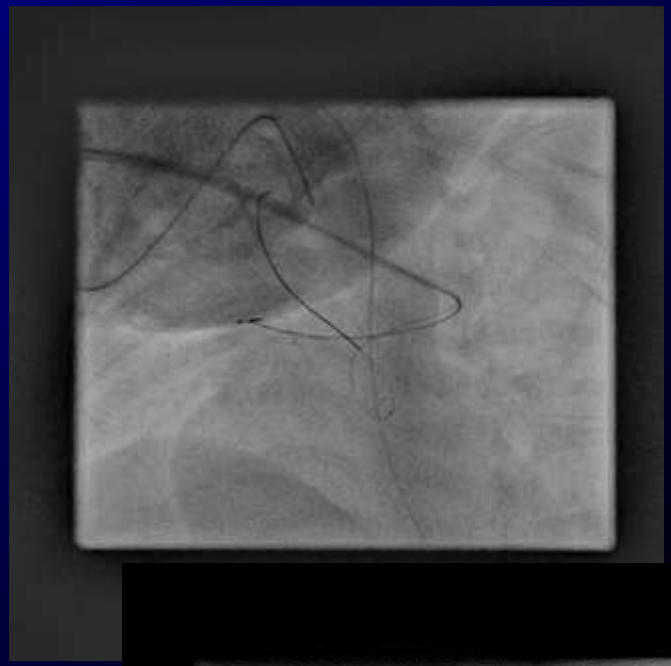
- Transvenous, transVSD. Use of Judkin's (R) guiding catheter (or XB guiding with distal tip removed)
- Trans-carotid approach – surgical cut down
- Trans-axillary (percutaneous)

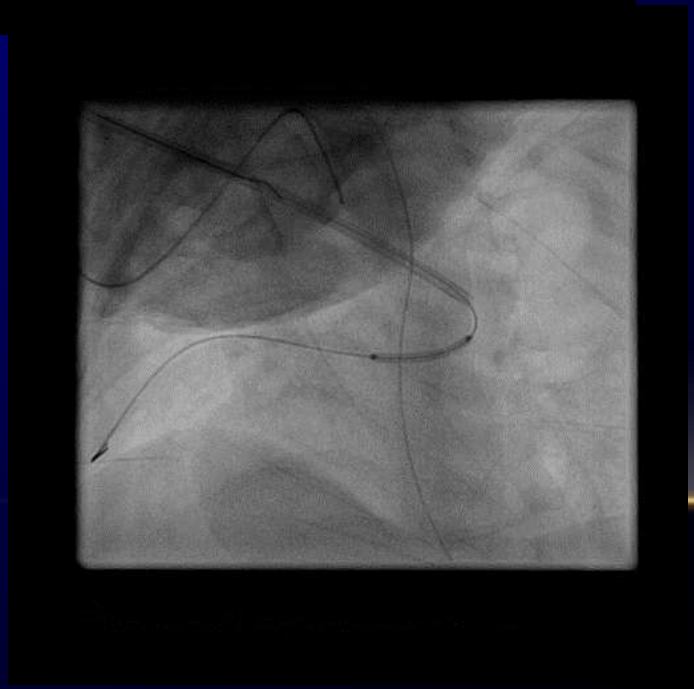
PDA stent via right axillary artery

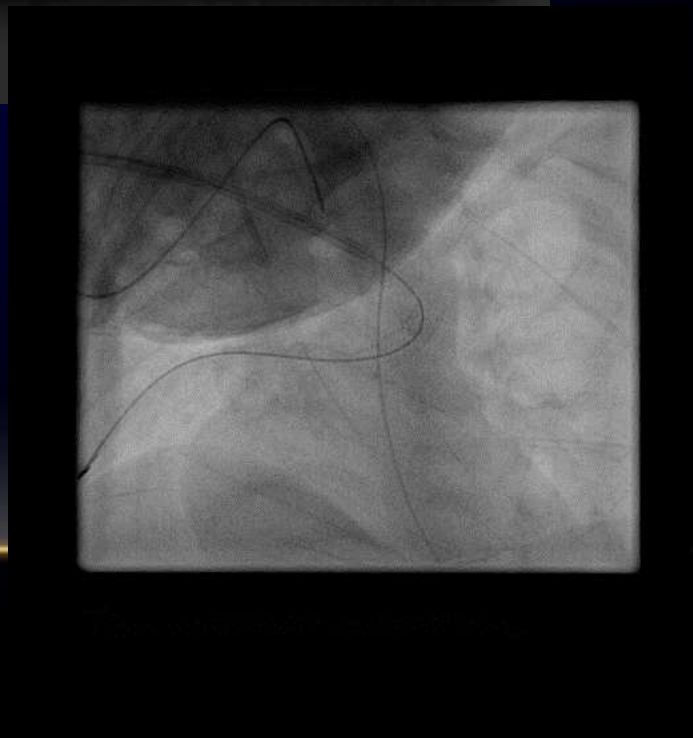


PDA stent via right axillary artery

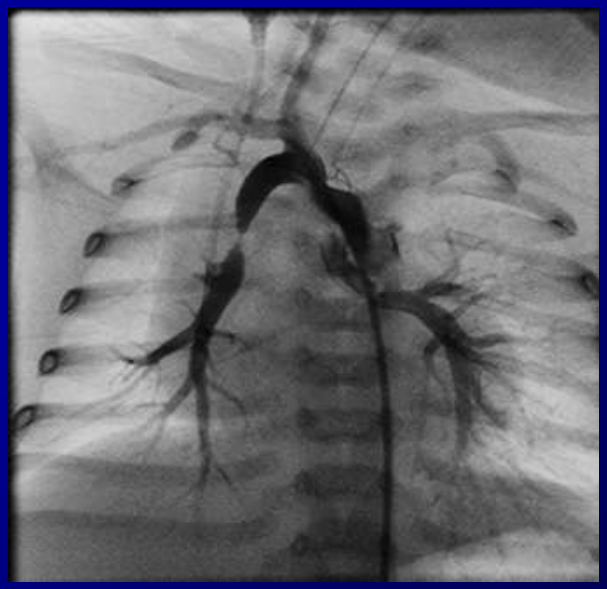






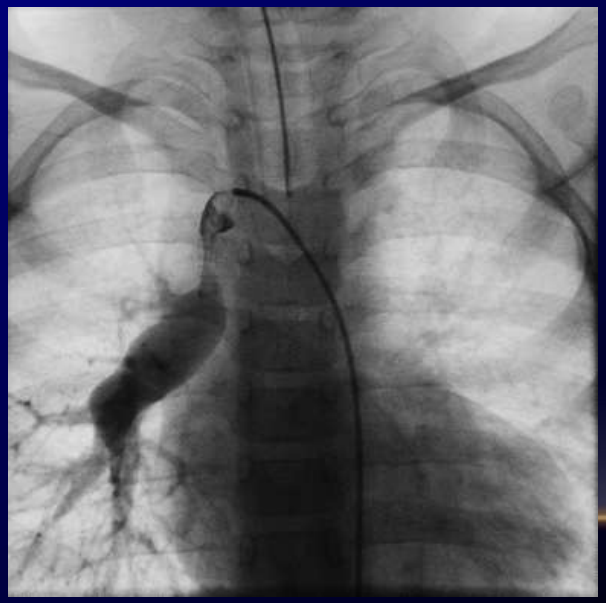


PDA stent in TOF-PA with ductus-related LPA stenosis



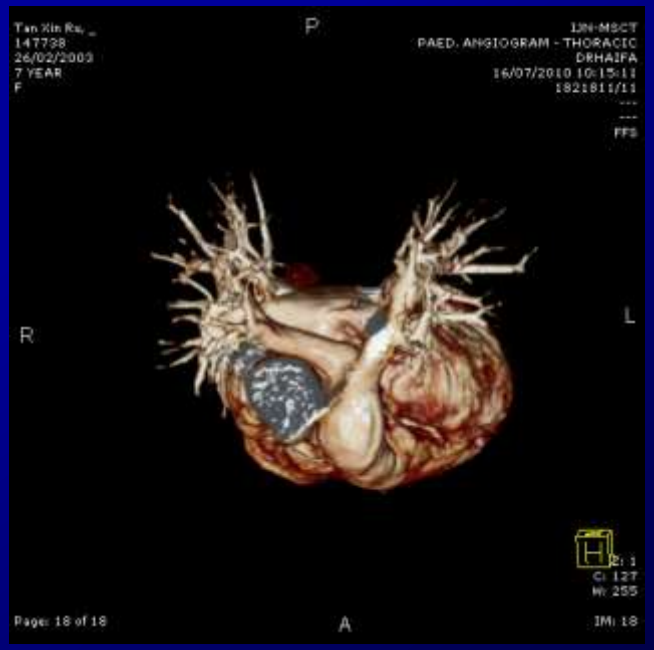
- Close follow up
- Good surgical team – early repair, excellent skills at repairing branch PA stenoses

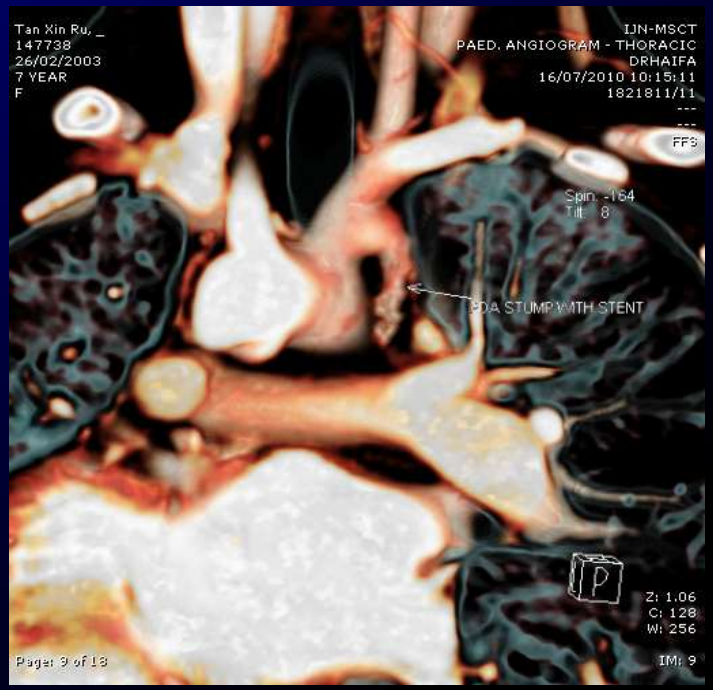
TOF-PA, bilateral PDA
2003 – PDA stent at 1 month,
left BT shunt at 5 months



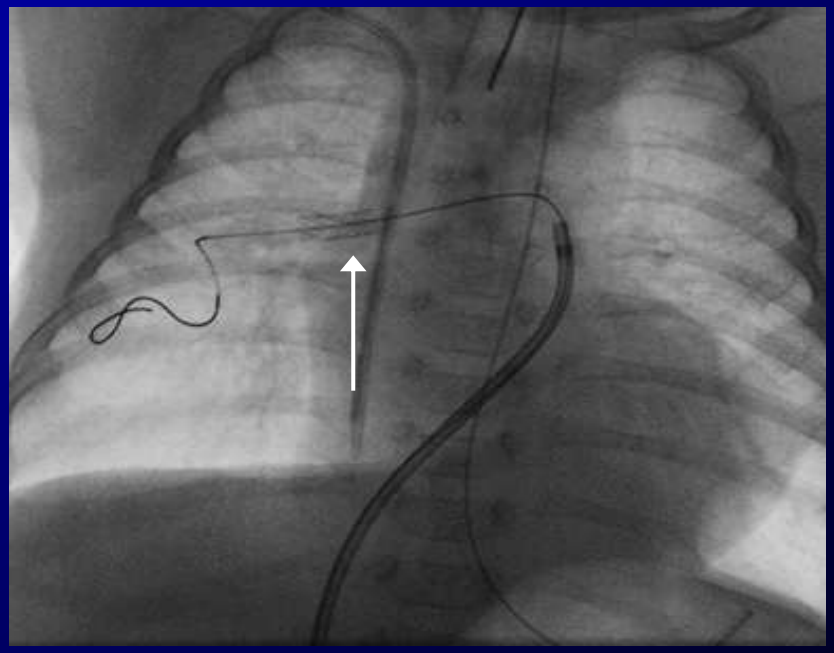
2006 (pre-Rastelli study)
PDA stent – good flow
Left BT shunt – almost occluded

2010 – evaluation by MSCT

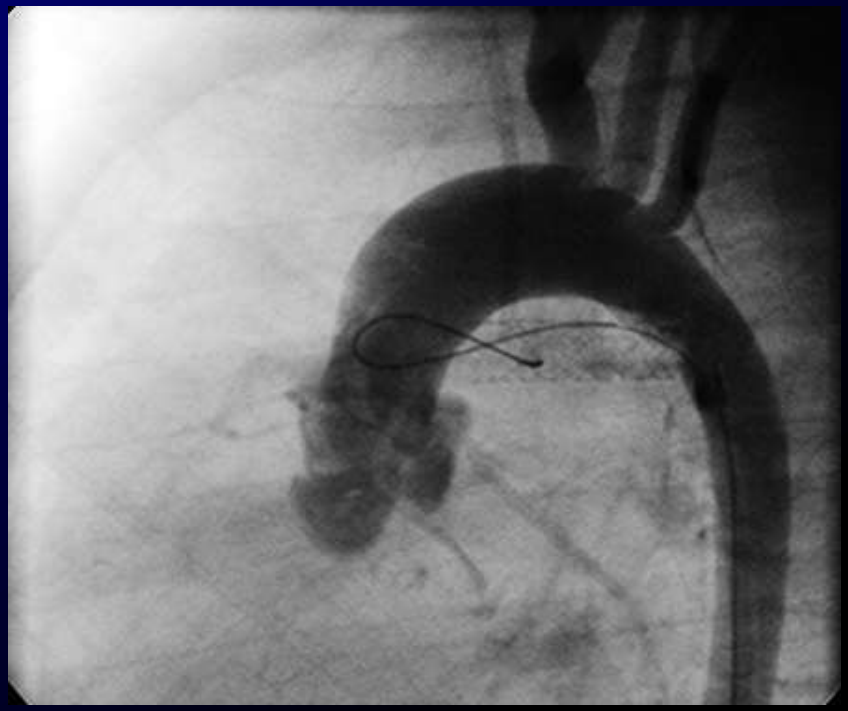




Major complications



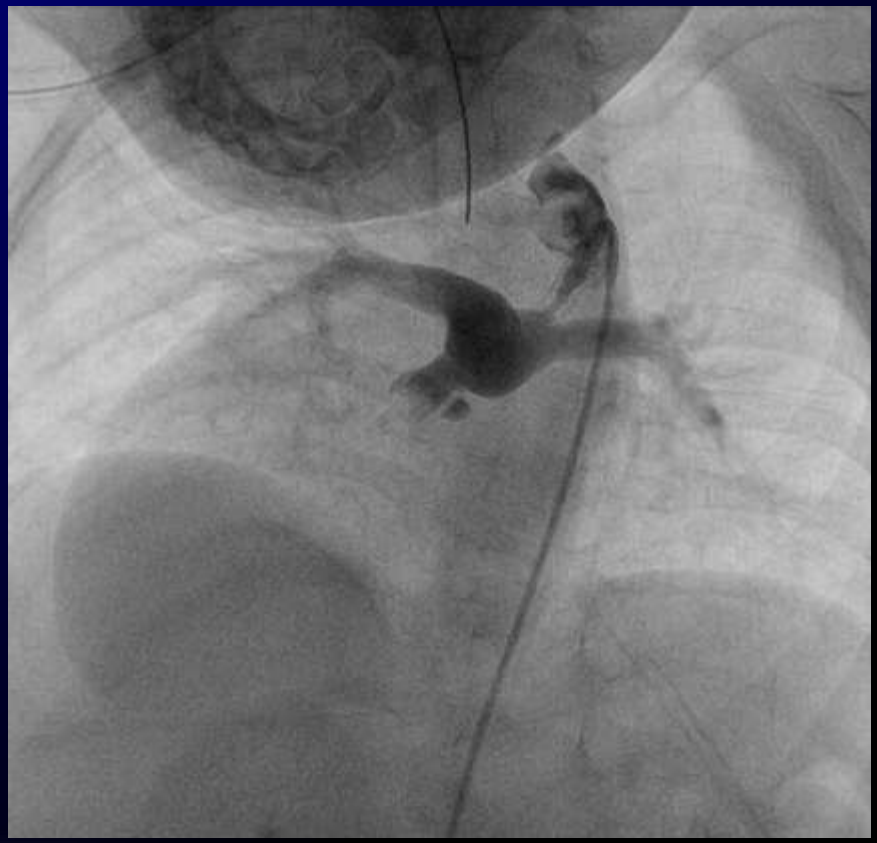
Stent migration



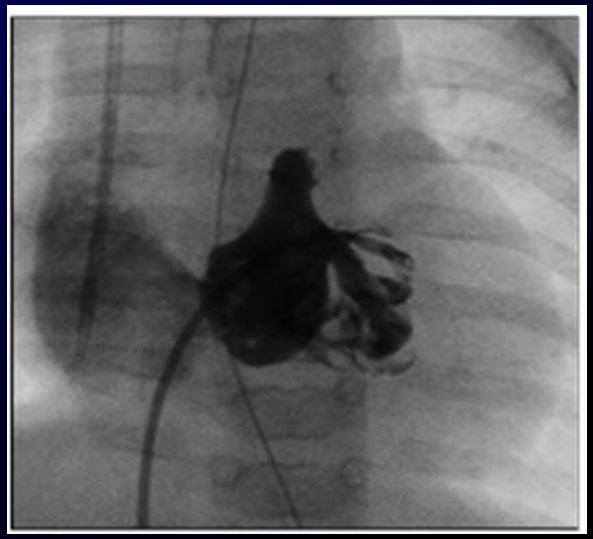
Acute thrombosis

Prevention: ? Pre treatment with Aspirin
heparization regime intra procedure

What happens to PDA stent with time?



Summary 1
PDA stenting… A tale of 2 pulmonary atresias



WHO …

- PA-IVS – bipartite RV, poorly developed apex ↓ RV cavity, ↓ RV compliance (RVH ++)

WHEN …

- Concomittant RF valvotomy and PDA stent

HOW …

- Via femoral artery route as most PDAs arise from distal arch

WHAT THEREAFTER …

- 2 ventricle or 1 ½ ventricle circulation depending of growth of RV

Summary 2
PDA stenting… A tale of 2 pulmonary atresias

WHO …

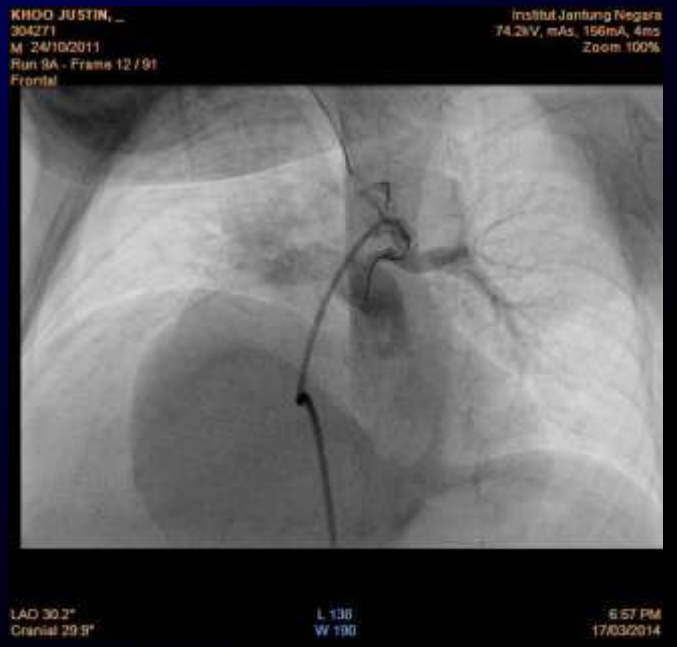
- TOF-PA variable PDA morphology
  Most common – PDA arising from underneath aortic arch and inserting onto LPA, ductus related LPA stenosis

WHEN …

- Late neonatal period (PDA begins to constrict)

HOW …

- Transvenous, transVSD
- Transcarotid (surgical cut down preferred)
- Transaxillary



WHAT THEREAFTER …

- Watch for LPA growth – close follow up, ? Needs rescue left BT shunt
- Surgical repair at 6 – 9 months + repair of LPA stenosis

Thank you