PA,VSD with MAPCAs

- Surgical Timing and Techniques -

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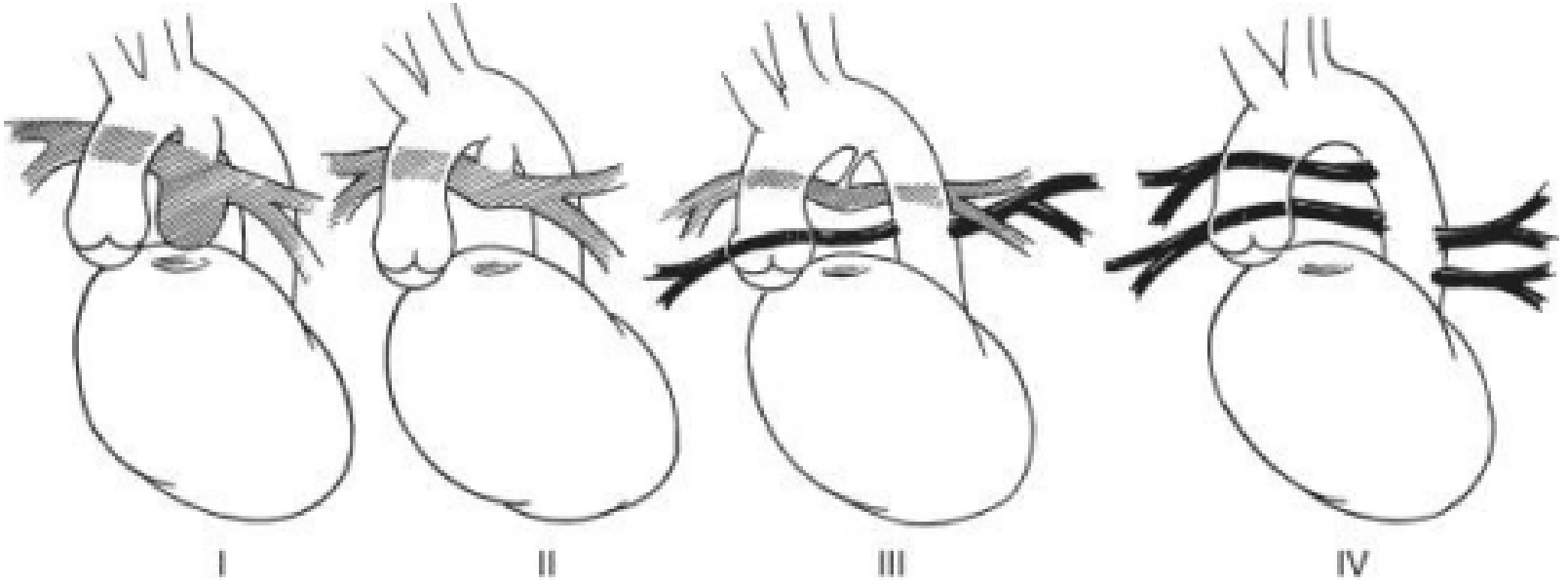
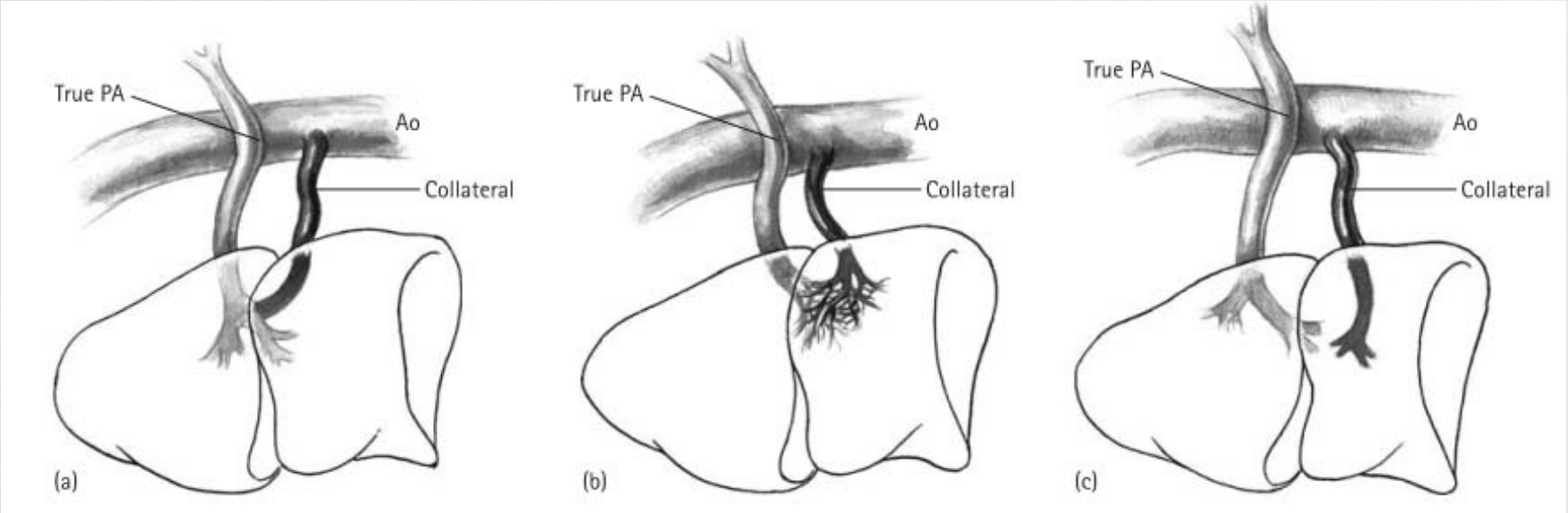
TOF Classification



Native Confluent Pulmonary Arteries **Major Aortopulmonary Collateral Arteries**

60~70% **20~30%** **5~10%**

Connection between MAPCA and true PA



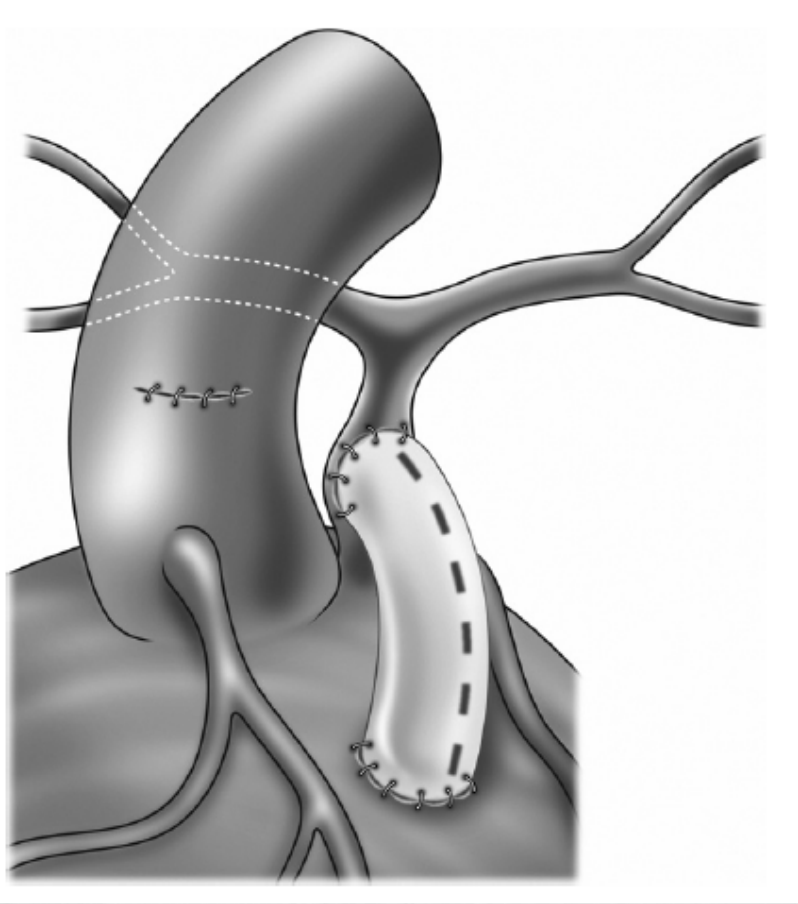
Timing

?

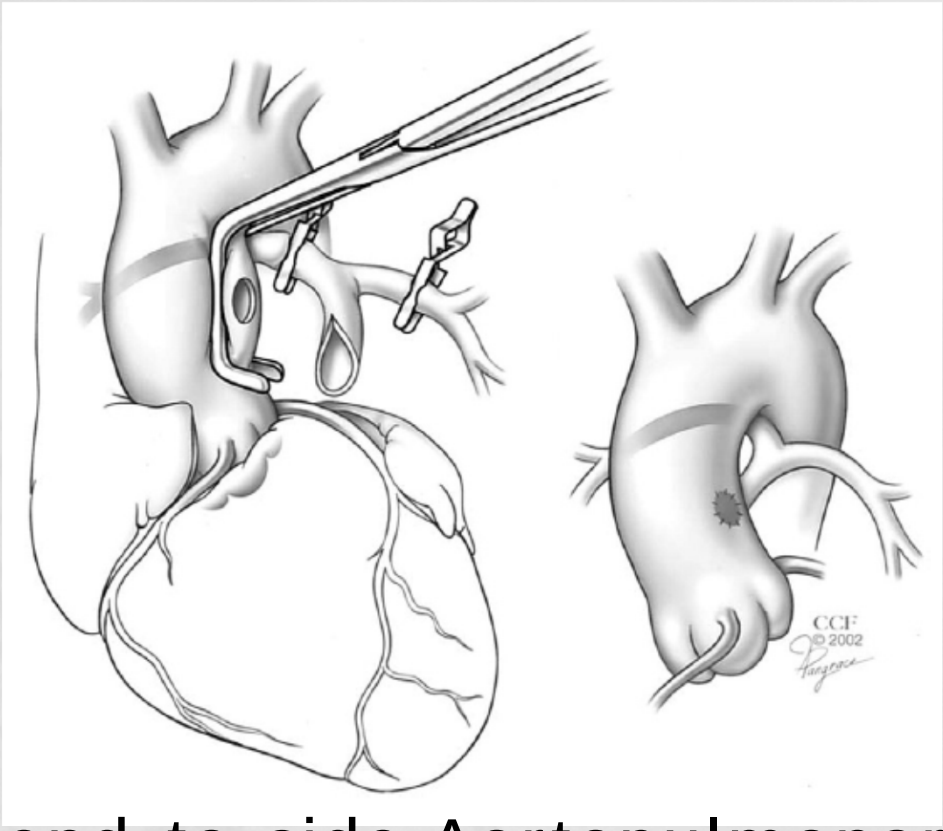
Surgical Strategy for PA with VSD, MAPCA

Central Confluent PA

RV-PA Conduit

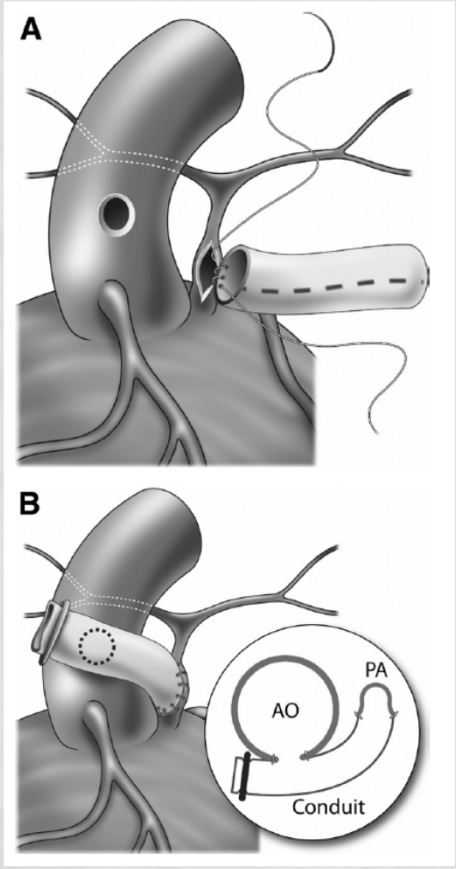


Melbourne Shunt



Central end-to-side Aortopulmonary shunt
Diminutive central pulmonary arteries

Modified Central Shunt



Pulmonary atresia with ventricular septal defects and major aortopulmonary collateral arteries: Unifocalization brings no long-term benefits

Yves d'Udekem, Nelson Alphonso, Martin A. Nørgaard, Andrew D. Cochrane, Leeanne E. Grigg, James L. Wilkinson and Christian P. Brizard

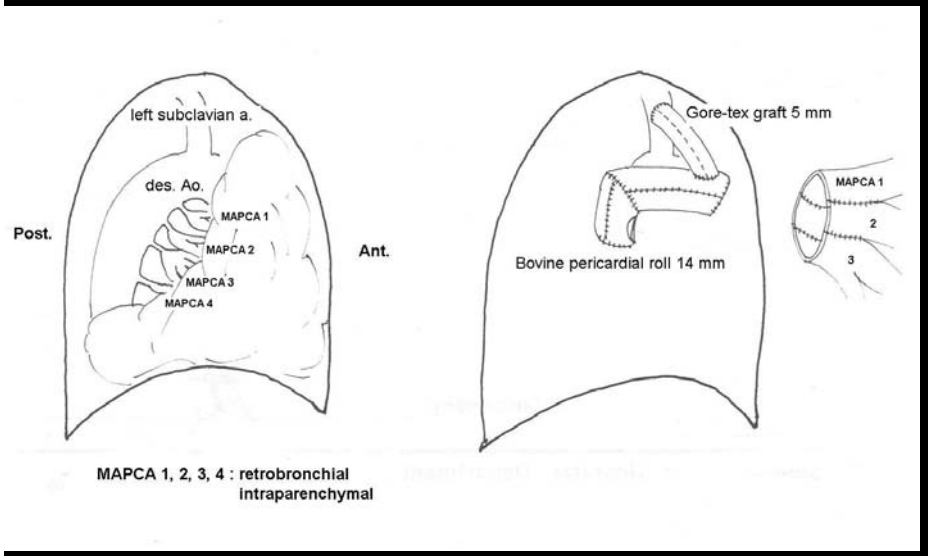
J Thorac Cardiovasc Surg 2005;130:1496-1502

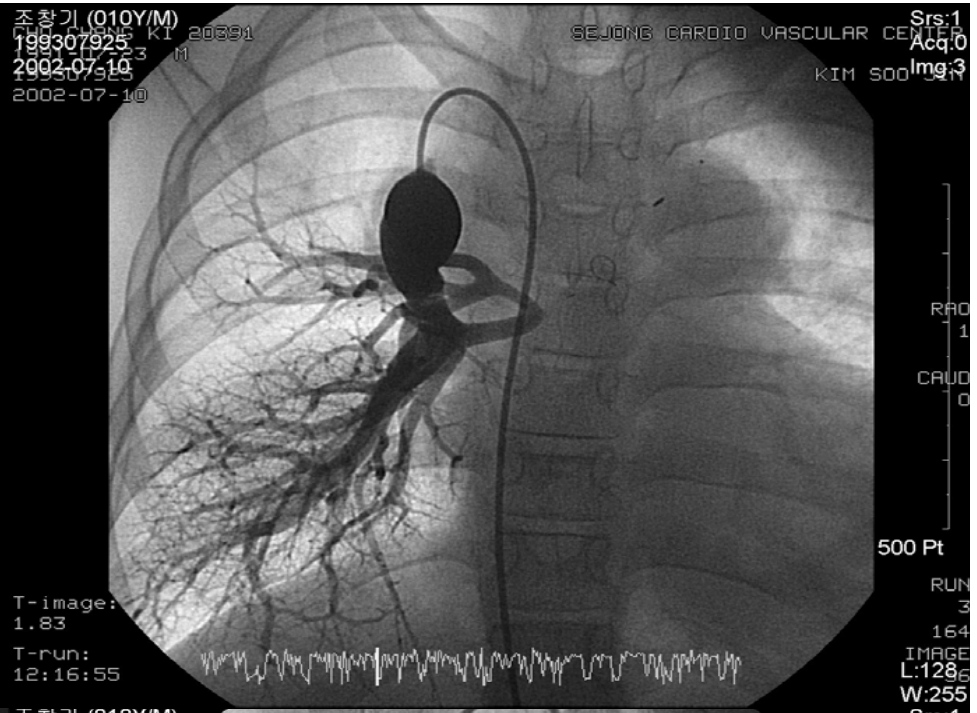
Late survival depends exclusively on the growth of the native pulmonary circulation.

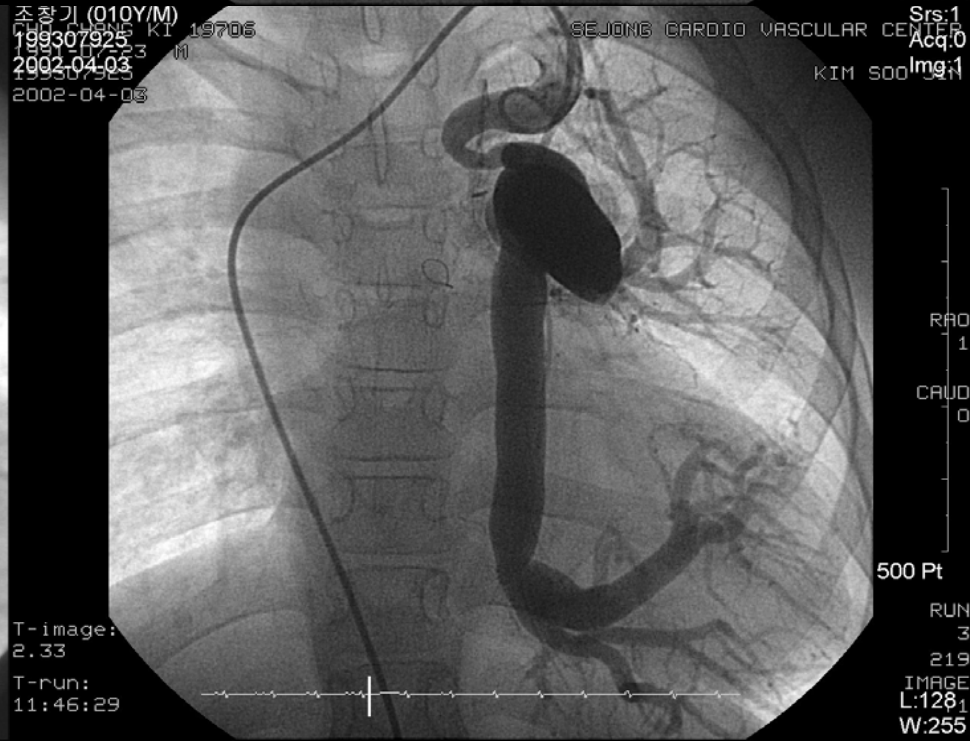
On angiography, central shunts promoted growth of central pulmonary arteries in all cases (29 patients). Sixty unifocalized major aortopulmonary collateral arteries were identified in 31 patients. After a mean of 3.2 ± 4 years, 26 thrombosed, and 12 presented with a stenosis of greater than 50%. Serial measurements of 29 major aortopulmonary collateral arteries showed no signs of growth (P = .25).

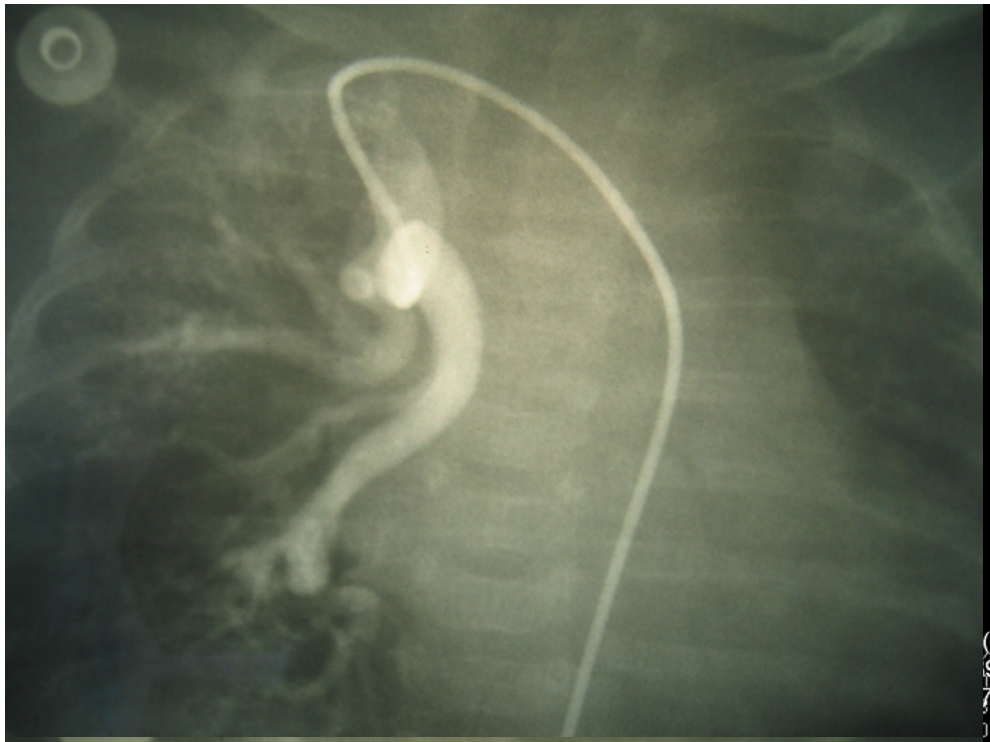
Unifocalization

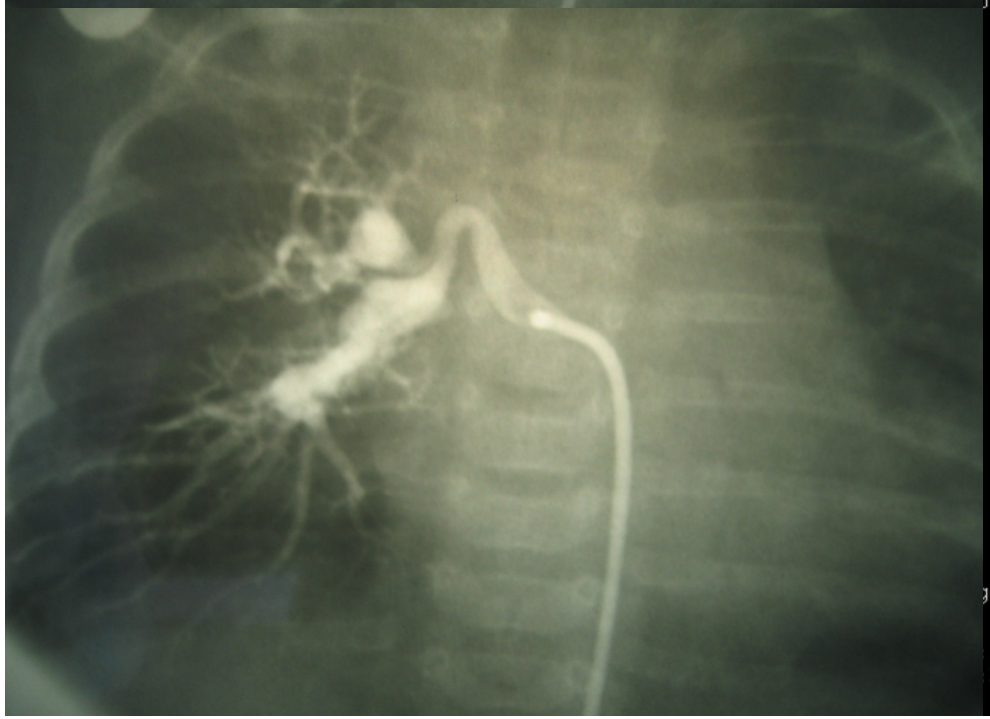
Unifocalization M / 10



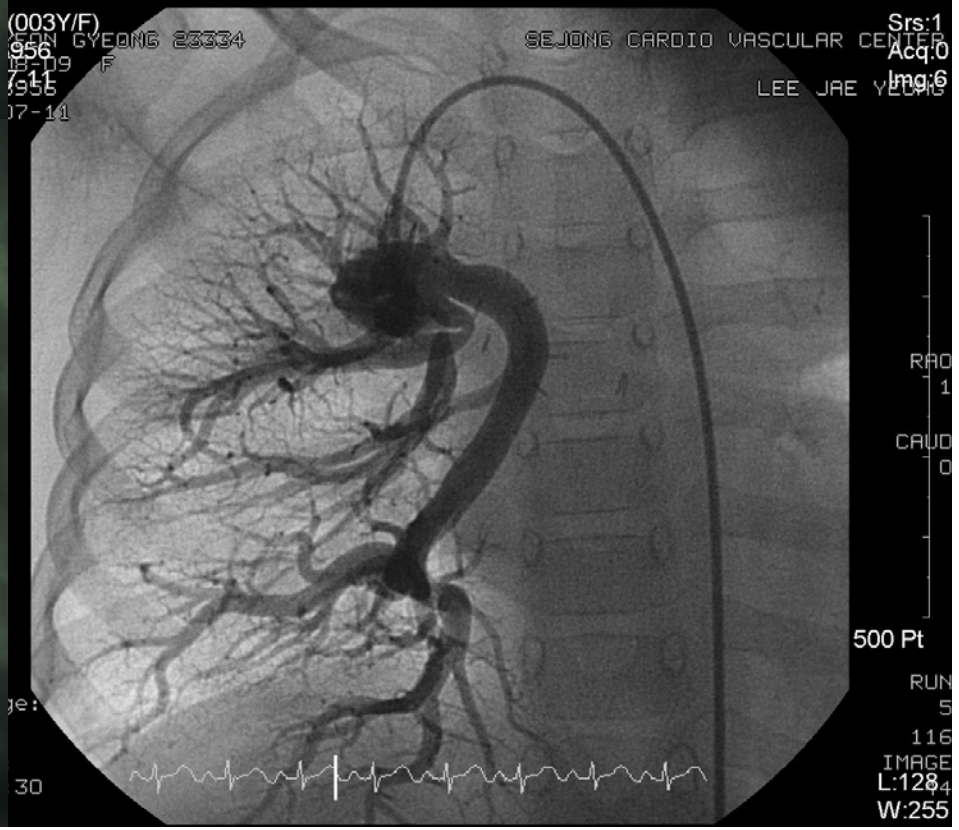








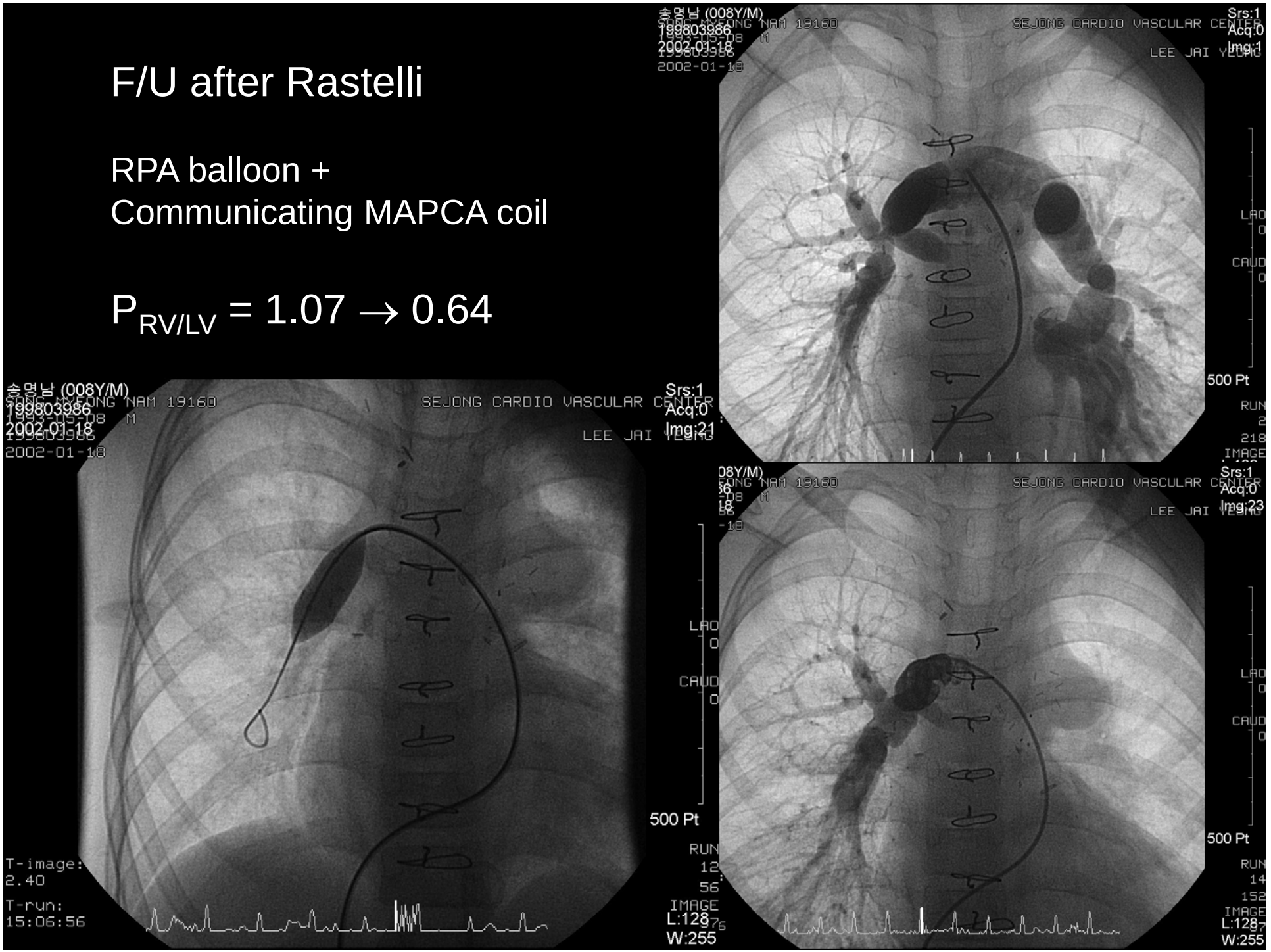
Rt. Unifocalization at 3 mo.
Native tissue-to-tissue
3 yr. later



F/U after Rastelli

RPA balloon +
Communicating MAPCA coil

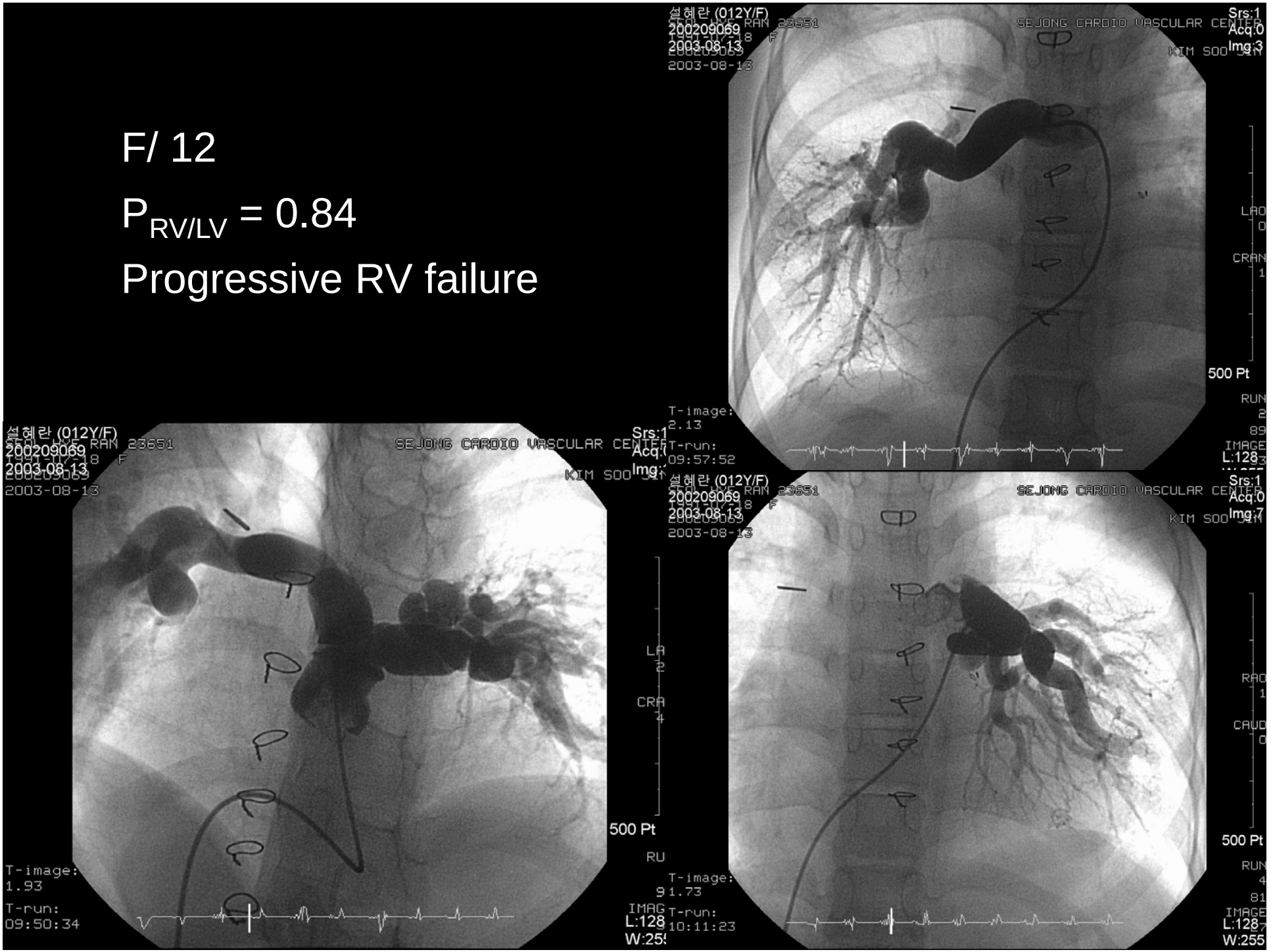
$P_{RV/LV} = 1.07 \rightarrow 0.64$



F/ 12

$P_{RV/LV}$ = 0.84

Progressive RV failure



Complete Repair : VSD closure
- Criteria -

- Central PA area ≥ 50% of predicted normal

  - Puga JTCS 1989;98(6):1028-9

- Predicted pRV/pLV ≤ 0.7, No MAPCAs remain

  More than 2/3 lung segments are centralized

  - by Iyer and Mee, ATS;1991:51:65-72

- Nakata Index > 150mm^2/m^2 BSA

  -by Metras, EJCTS 2001;20:590-6

- TNPAI ≥ 200 mm^2/m^2 *- by Hanley, JTCS 1997;113(5);858-66*

- 15 out of 20 bronchopulmonary segments(1 & 1/2 lungs) are connected to confluent pulmonary artery

  - by Baker, 2002

Personal Experience

- From 2004 to 2011
- 16 patients (male=8, female=8)
- Mean F/U duration
  - 47.7 ± 29.6 months

Mortality cases

- 3 mortalities
  - Cardiac related death
    - 1 patient (F/40, Progressive RV failure)
  - Non-cardiac related death
    - 2 patient
      - CR (+) group
        - 1 patients-pneumonia
      - CR(-) group
        - Severe lung disease (CCAM)

Mortality 1

#1. Heart : PA, MAPCA, VSD

-1 mo: unifocalization, PA banding

-9 mo: unifocalization (Rt.MAPCA to RPA)

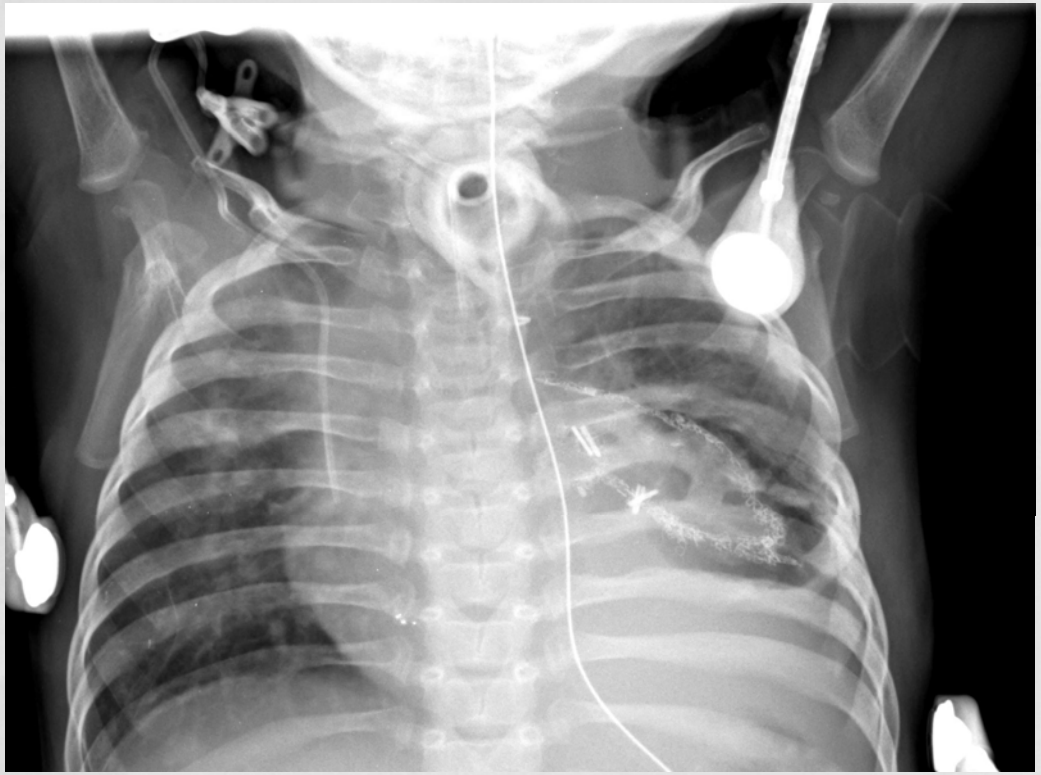
#2. Lung : CCAM (type 2, emphysematous change)

-2 mo: lung volume reduction surgery

-9 mo: LLL lobectomy

- Tracheostomy

- Persistent respiratory difficulty
- Prolonged ventilator care
- Desaturation and bradycardia
- 13mo , expired



Mortlity 2

- PA, VSD, MAPCA

  s/p 외부병원

  1 year-old Lt. mod. B-T shunt (5 mm)
  3 year-old Rt. mod. B-T shunt (6 mm) + unifocalization

- M/13 / 25 kg 2008.3.11. (aortic dilatation (46 mm) with AR
  -Ascending aorta & hemiarch replacement (graft 28 mm)
  -Complete repair : Rastelli operation
  - Carpentier-Edwards Valved Conduit 20 mm
  - VSD closure

#1. Respiration complication

- 1) underlying restrictive lung disease d/t scoliosis
  2) long-term ventilator care
  - pneumoniae
  - UTI (fungus)

- Hypoxic brain damage after respiratory arrest
  - Bed ridden state
  - Prolonged ventilator care

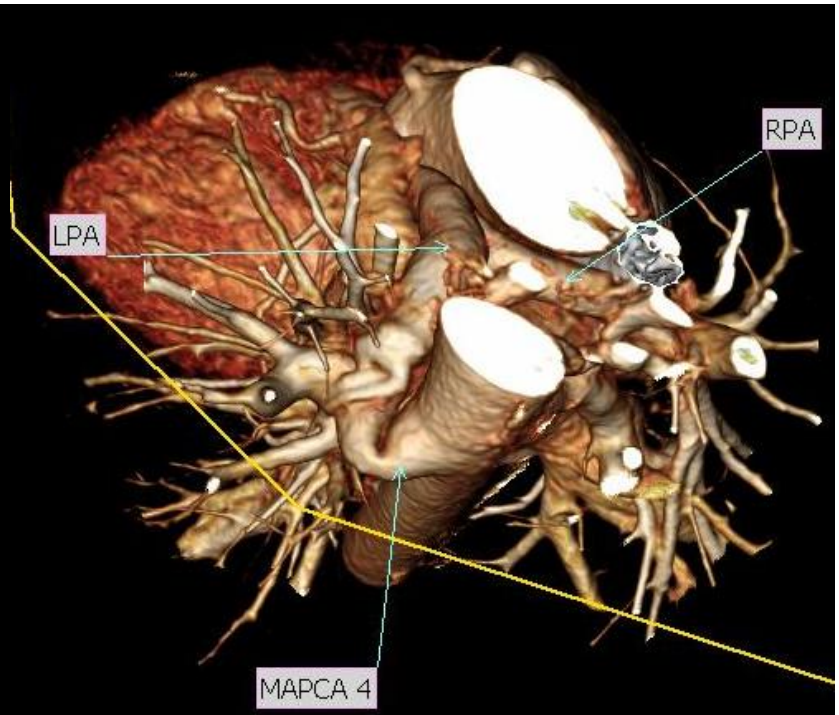
- Mortality : postop. 2 year later

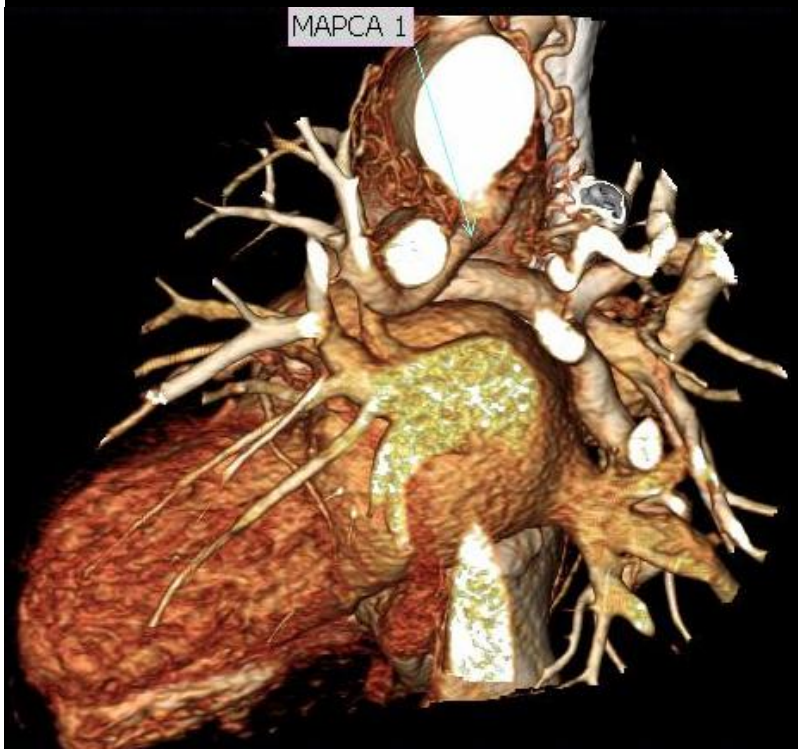
CASE 1

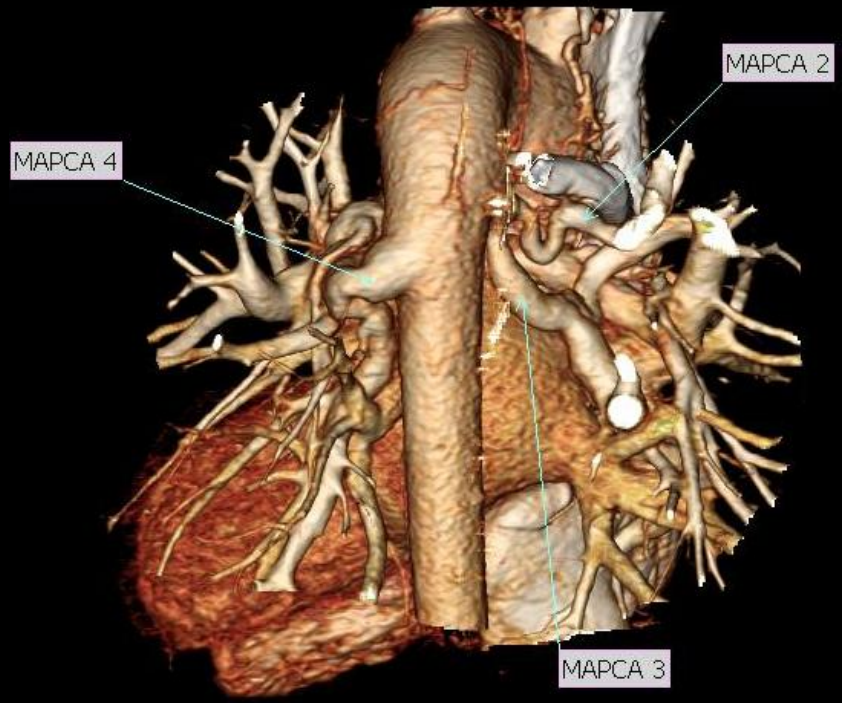
- F/27
- Before Marriage

PA/VSD/MAPCA

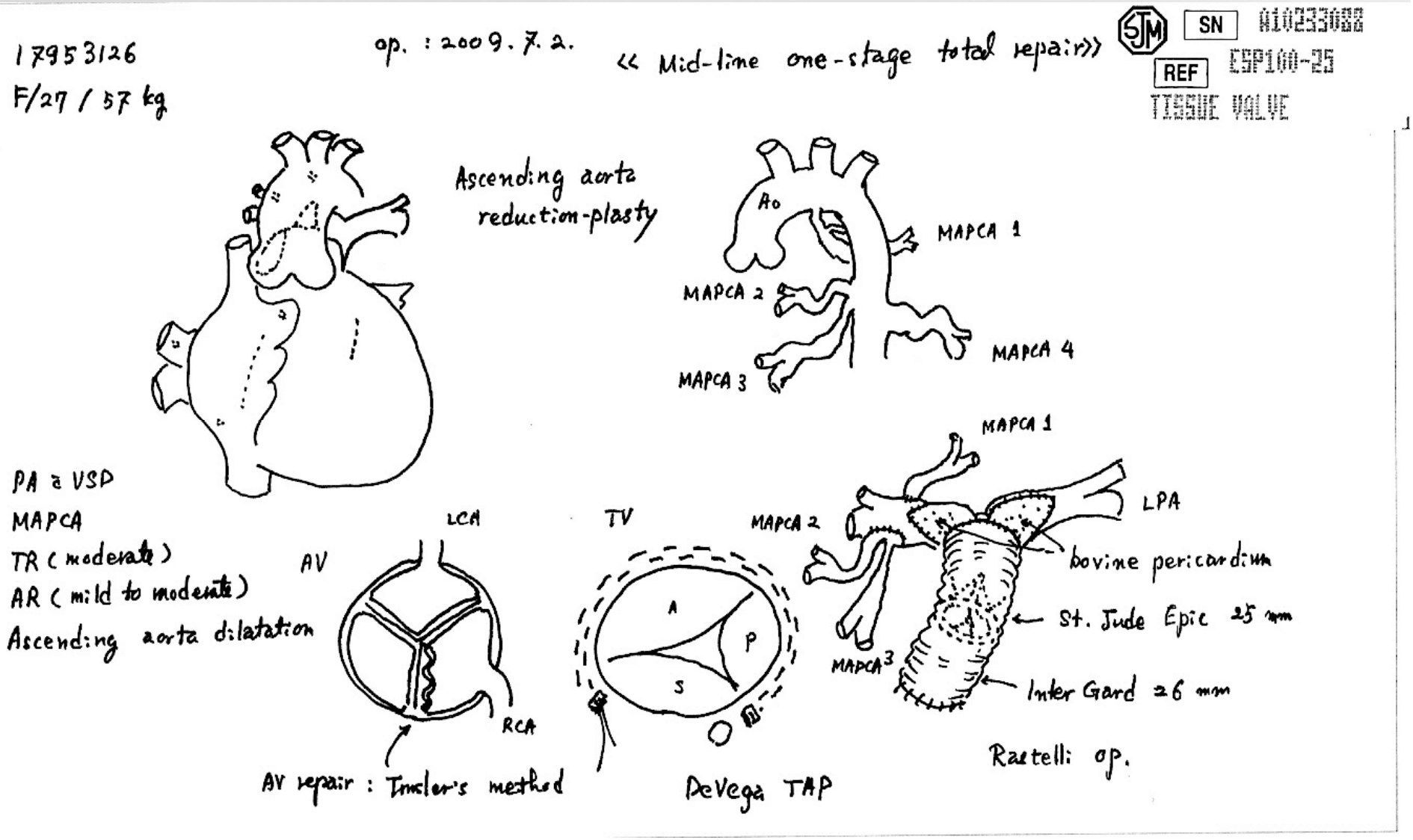
F / 27
Aortic root dilatation
AR, TR : moderate

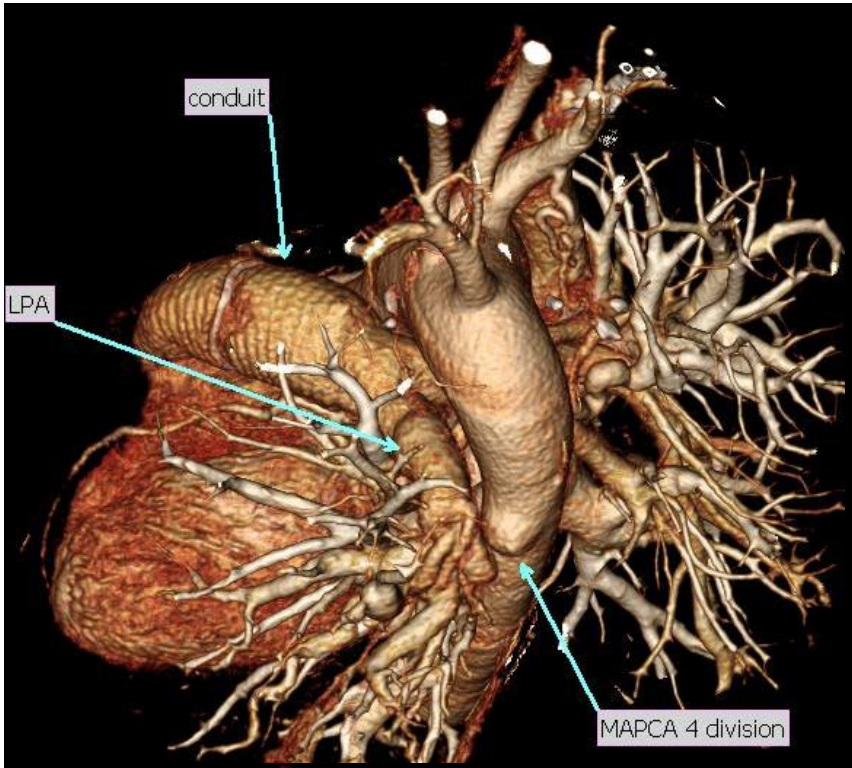


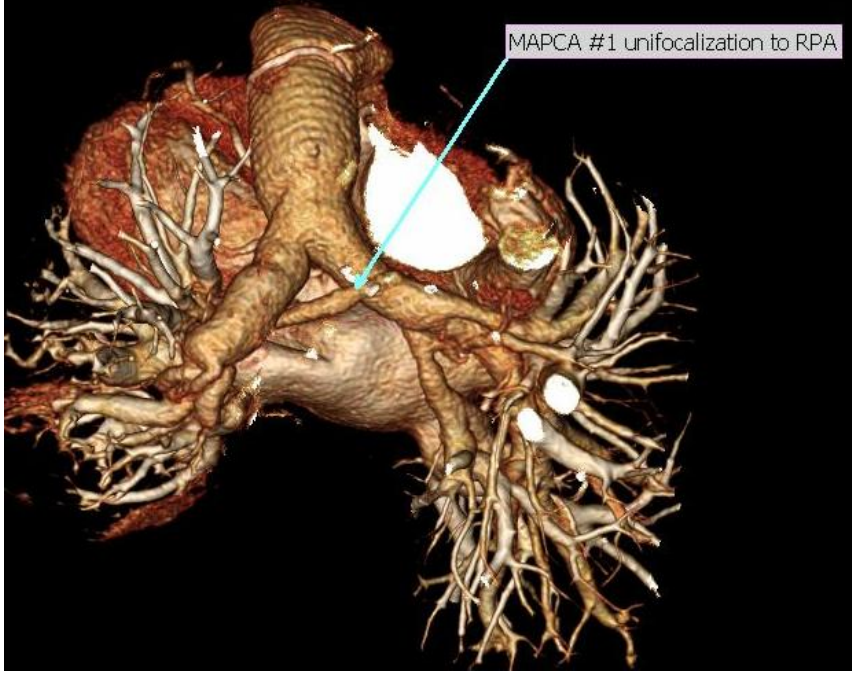




Midline One-stage Complete Repair

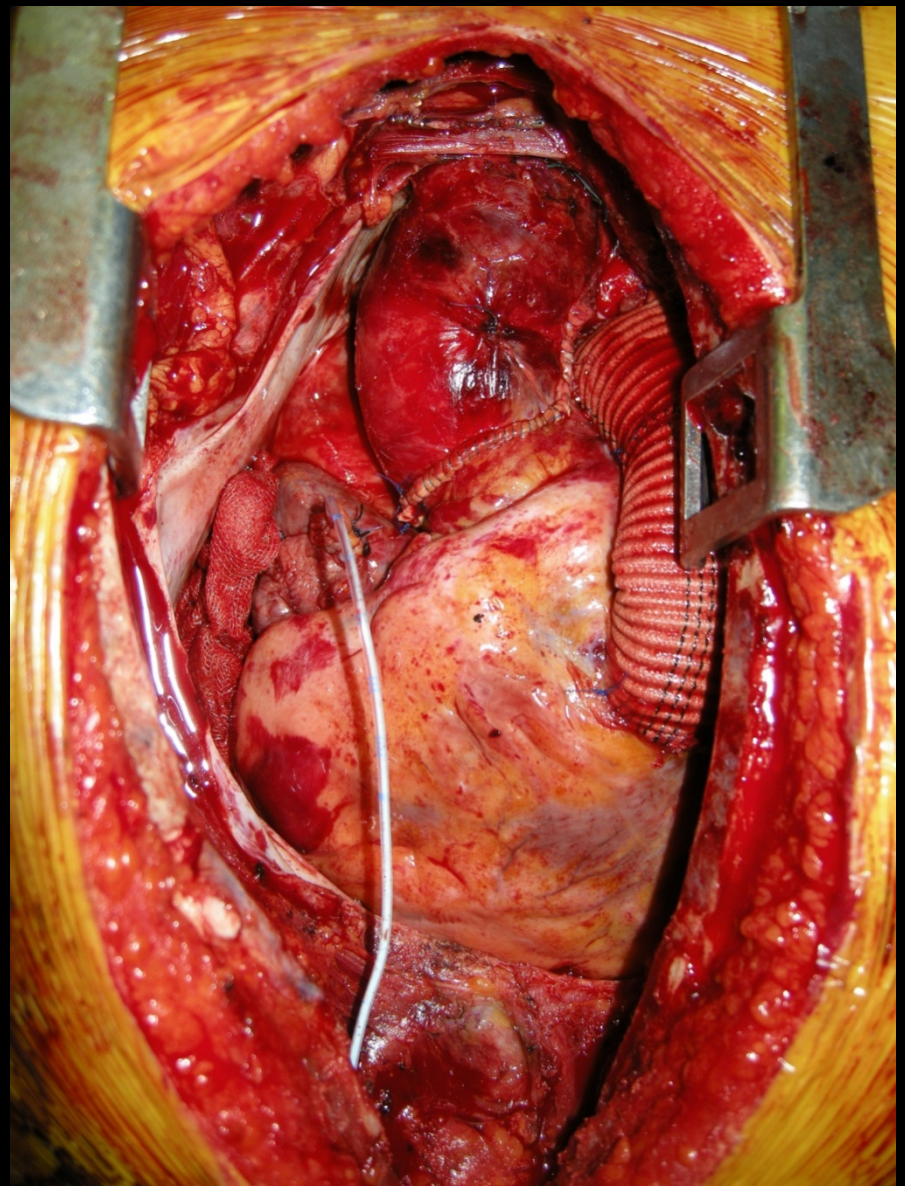






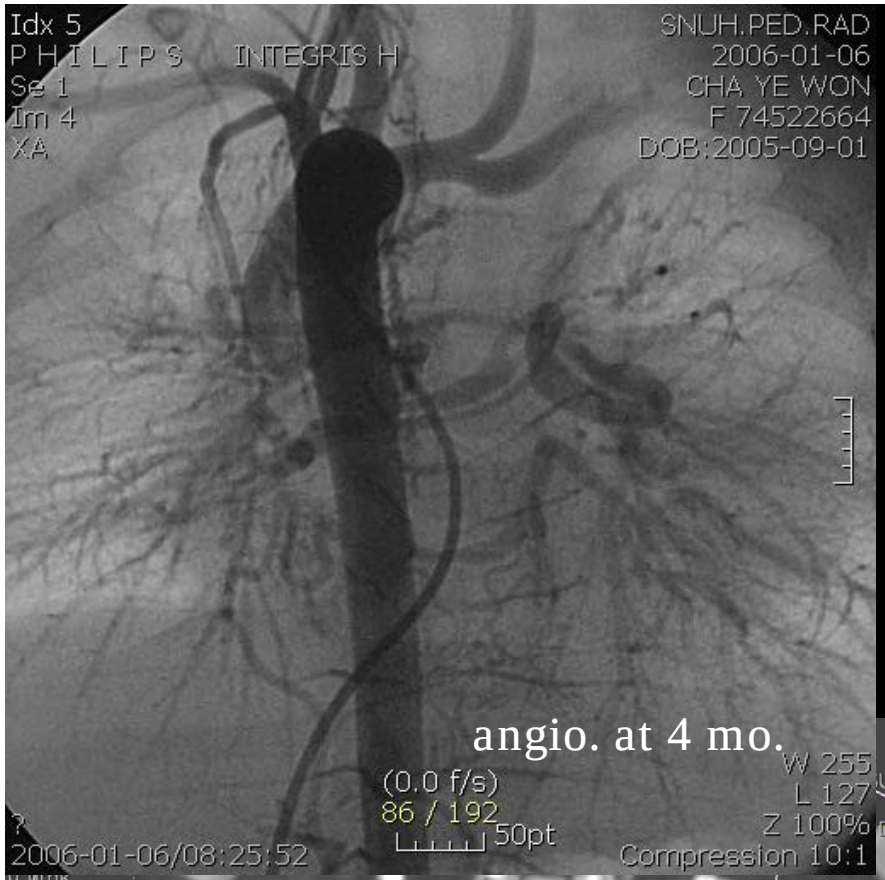
Midline One-stage Complete Repair

F / 27 Postop. $P_{RV/LV} = 0.65$

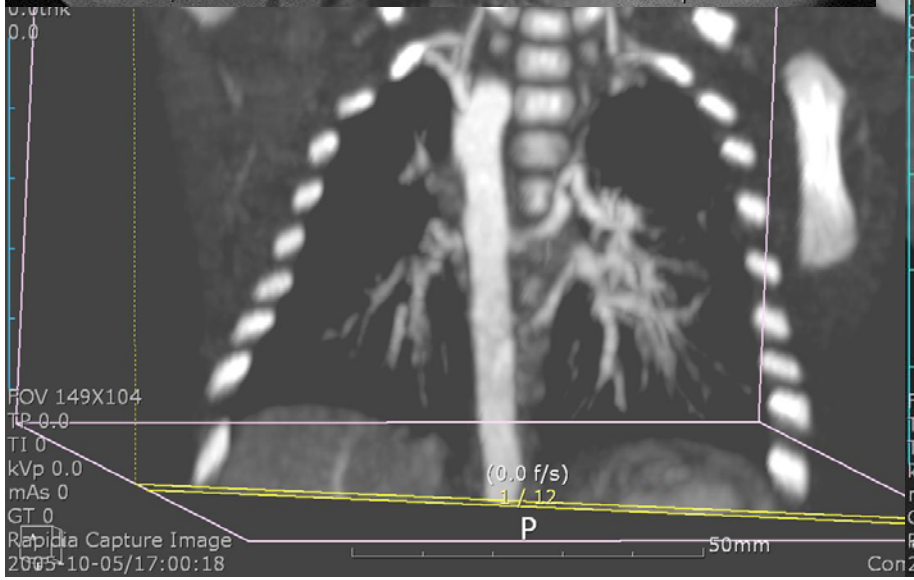


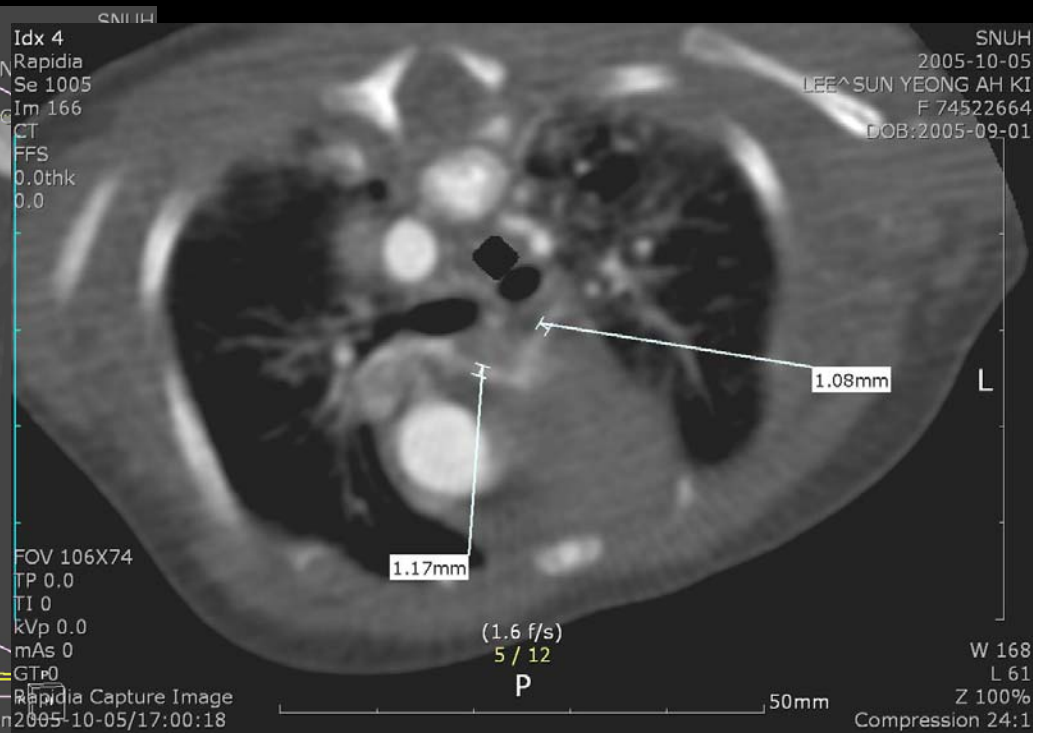
Case 2

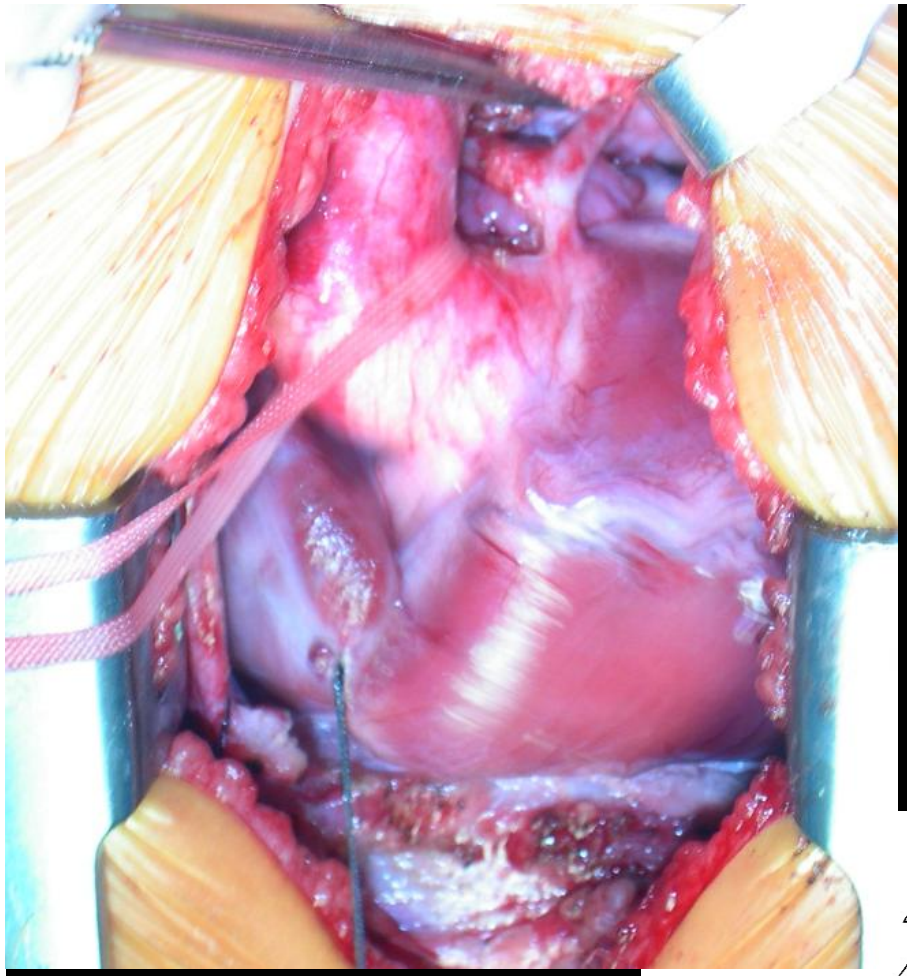
F / 33 +3 wk
Birth wt. = 1.6 kg
Confluent PA : 1 mm



angio. at 4 mo.

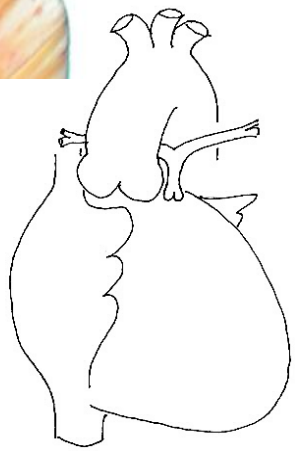


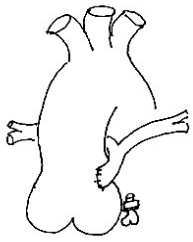




1st operation
4 mo/5.4kg

Melbourne shunt



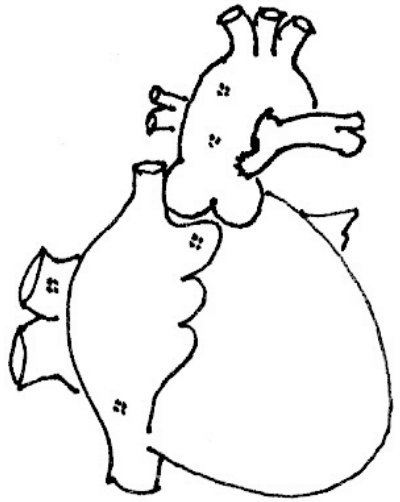


confluent central PA
reimplantation to aorta

2nd operation
9 mo/6.8 kg

Palliative RV to PA conduit interposition
—Goretex tube graft 8 mm, Both PA angioplasty

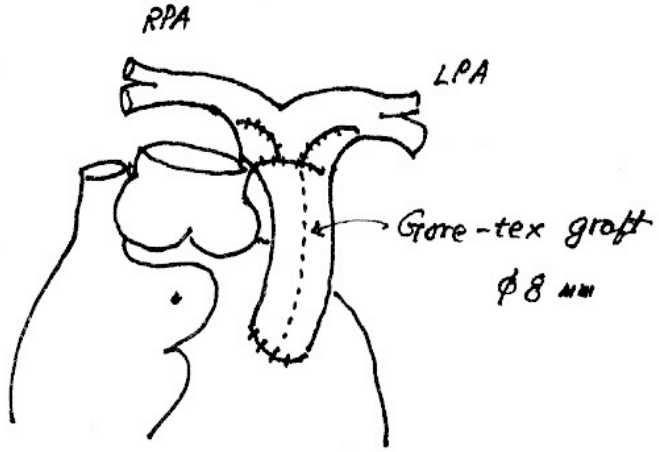
PA, VSD, MAPCA (multiple, hypoplasia)



initially rudimentary PA (φ1 mm)

s/p 2006. 1. 12. MPA reimplantation to aorta

→ well growing PA

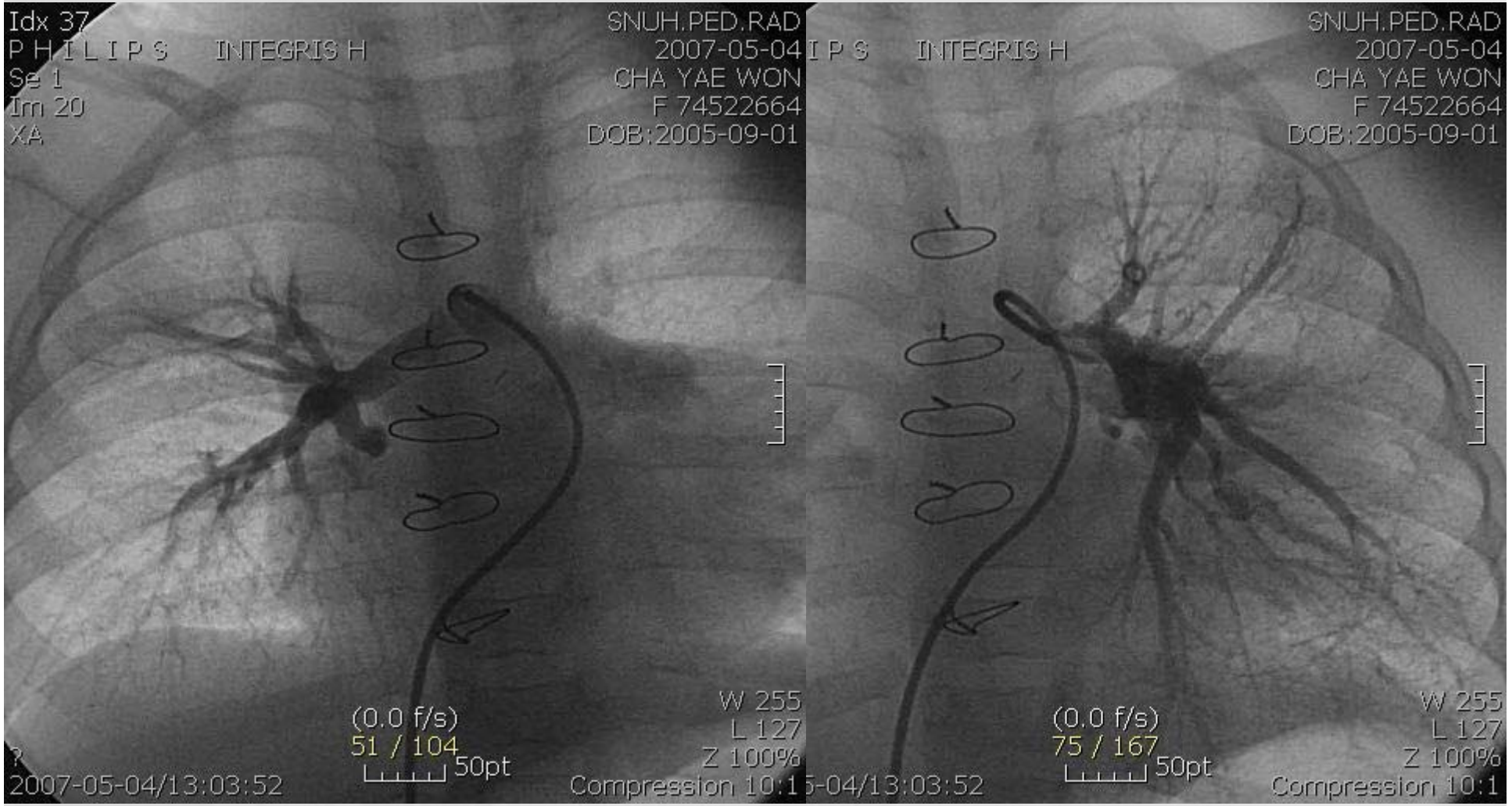


Palliative RVOT reconstruction
(RV-PA conduit)

12month

- Ballon angioplasty of LPA and distal RPA

F / 1 yr. 8 mo.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50pt
W 255
L 127
Z 100%
Compression 10:1
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DOB:2005-09-01
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75 / 167
50pt
W 255
L 127
Z 100%
Compression 10:1
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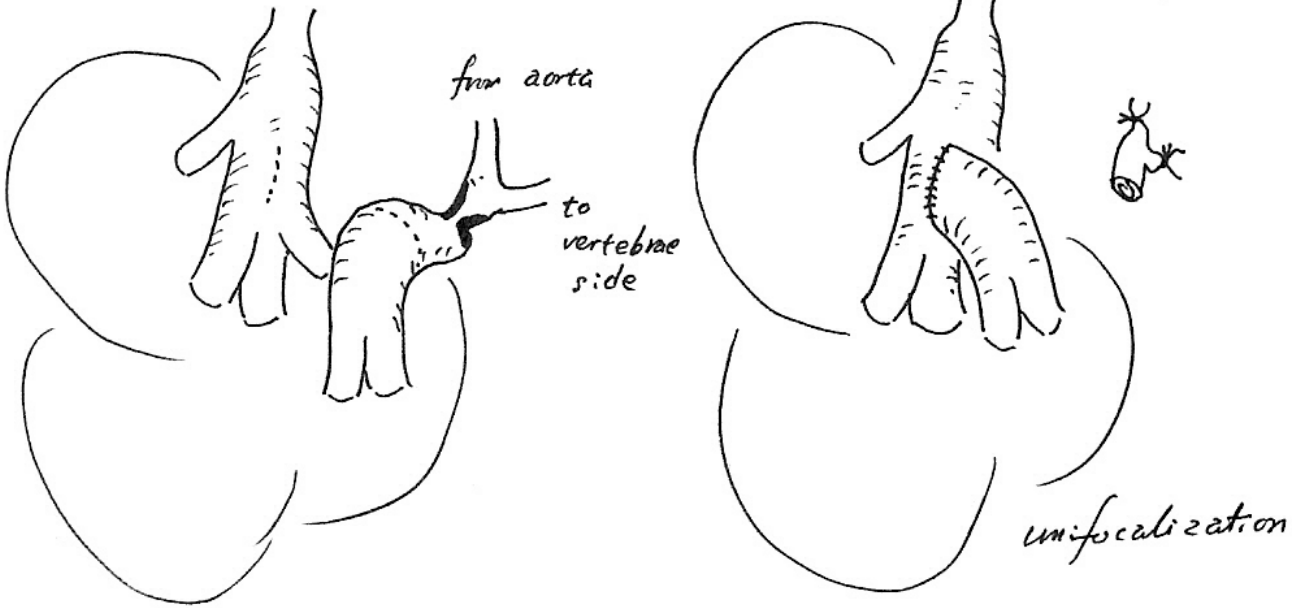
3rd operation
21 mo/9.9 kg

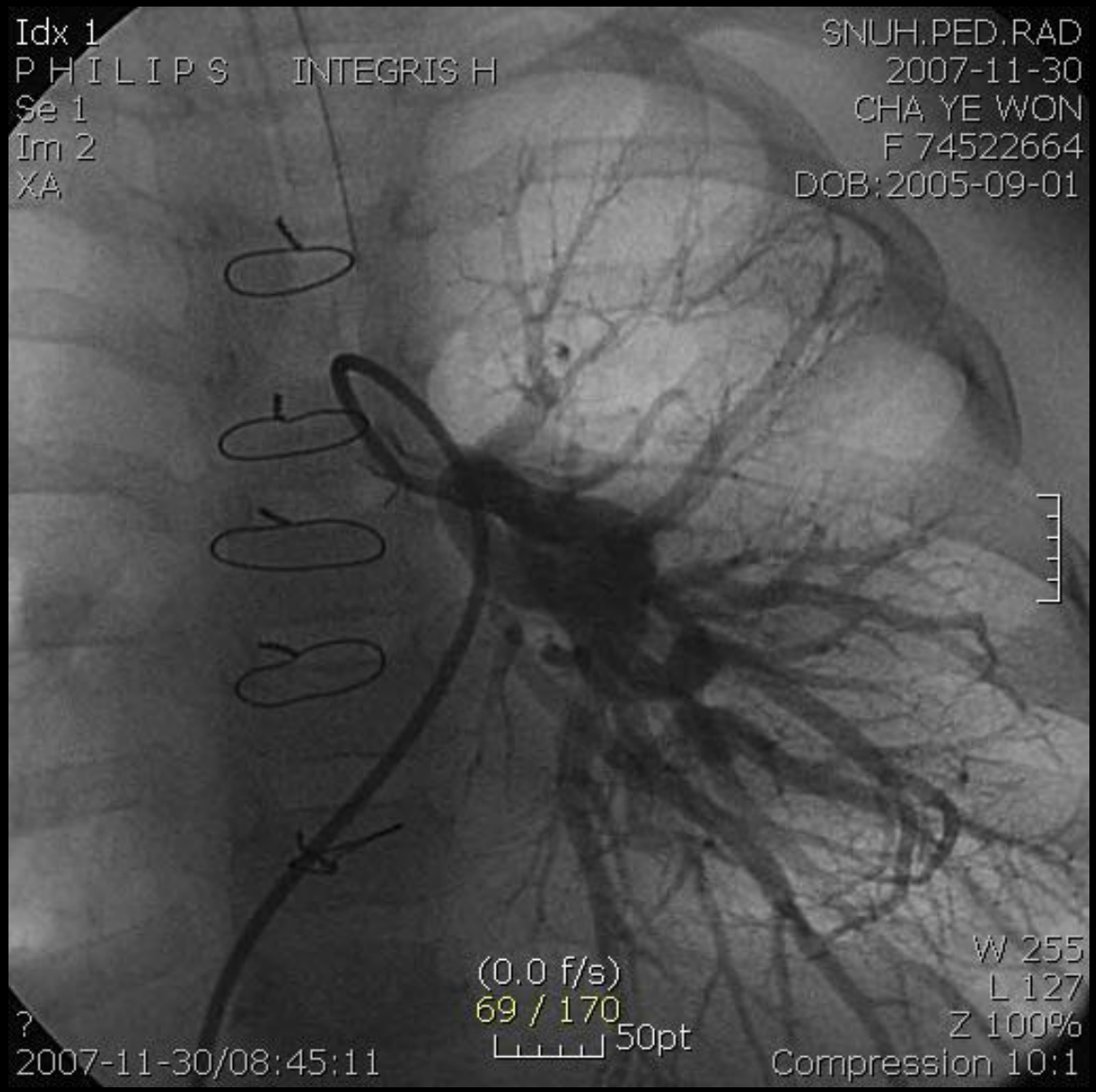
Unifocalization of MAPCA, left

PA, VSD, MAPCA
Rudimentary confluent PA (1 mm)

s/p 2006. 1. 12. MPA reimplantation to aorta
2006. 6. 12. Palliative RV-to-PA conduit
(Gore-tex tube graft 8 mm)
Both PA patch angioplasty



F / 2 yr. 2 mo. F/U Angio.

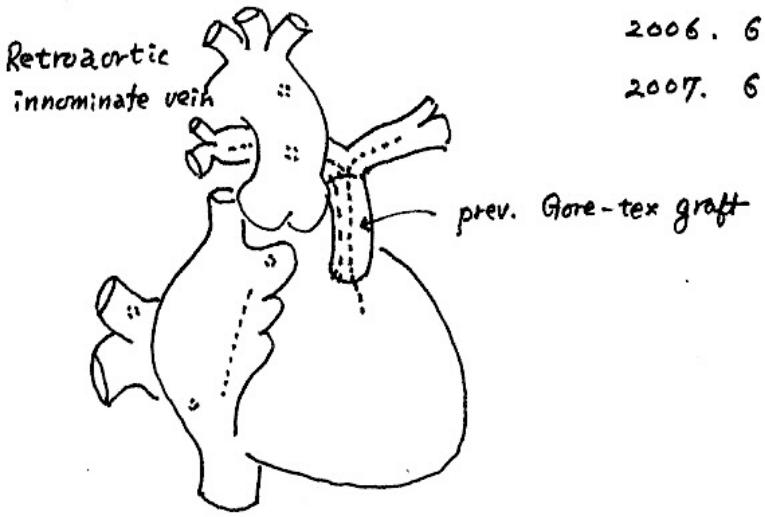


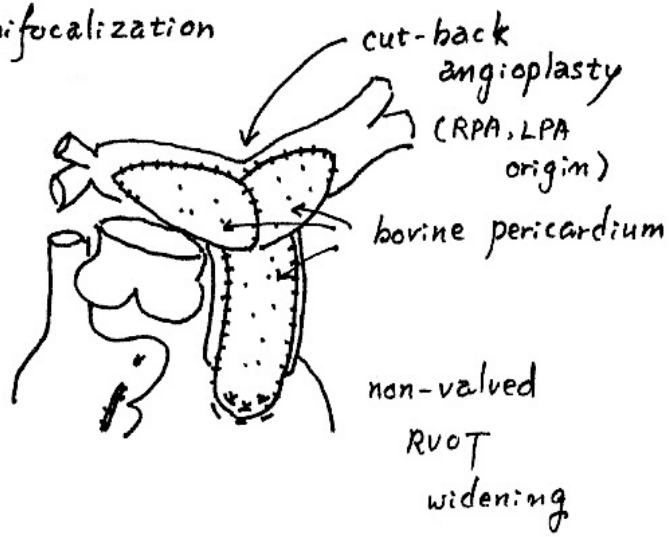
26 month

- Balloon angioplasty of proximal LPA

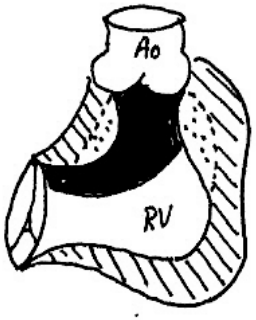
4th operation
30 mo/10.7 kg

s/p Preterm birth 33+3 wk, 1.6 kg
2006. 1. 12. MPA reimplantation to aorta
2006. 6. 12. Palliative RV-PA conduit
2007. 6. 4. Left unifocalization



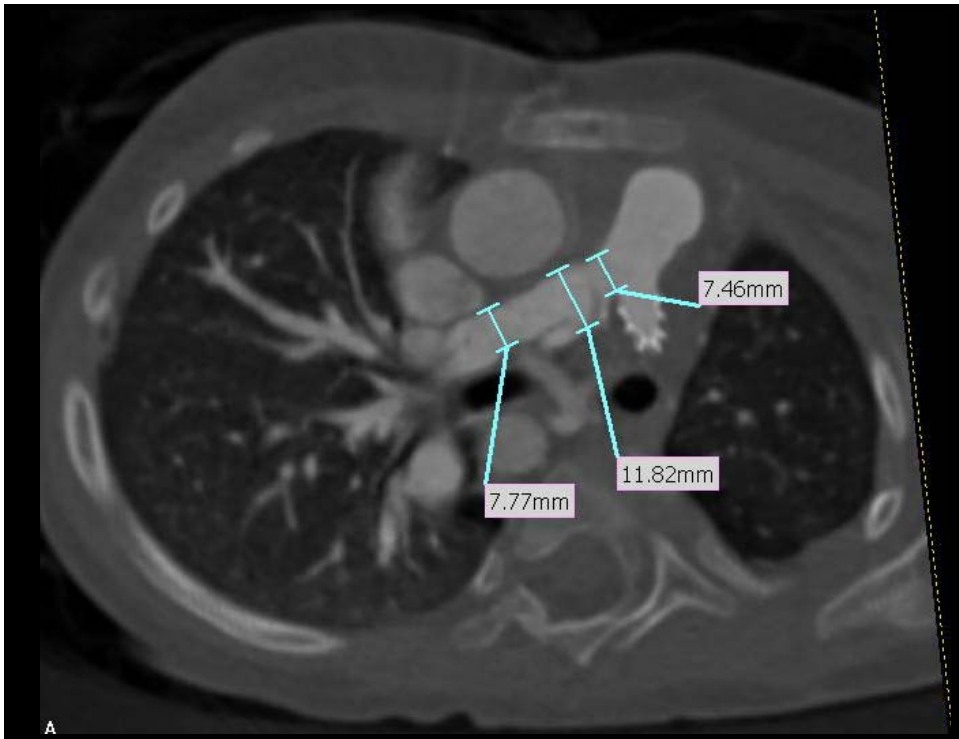


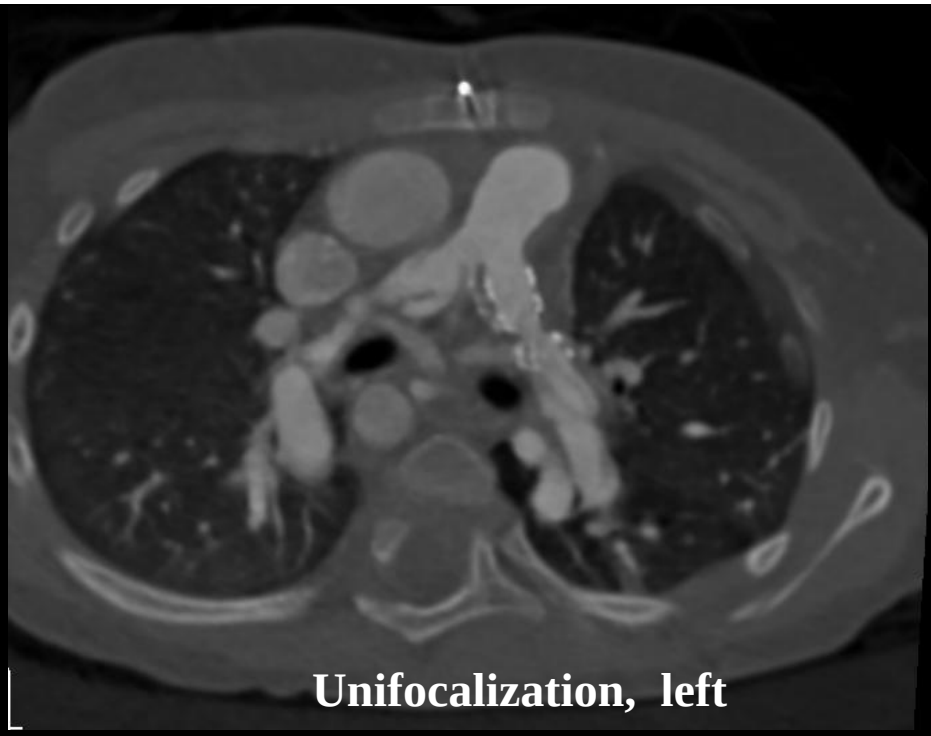
Total Repair
- **—Both PA patch angioplasty**
- **—VSD patch baffling, RVOT patch widening**
- **—RV infundibular muscle wide excision**
- **—PFO primary closure**



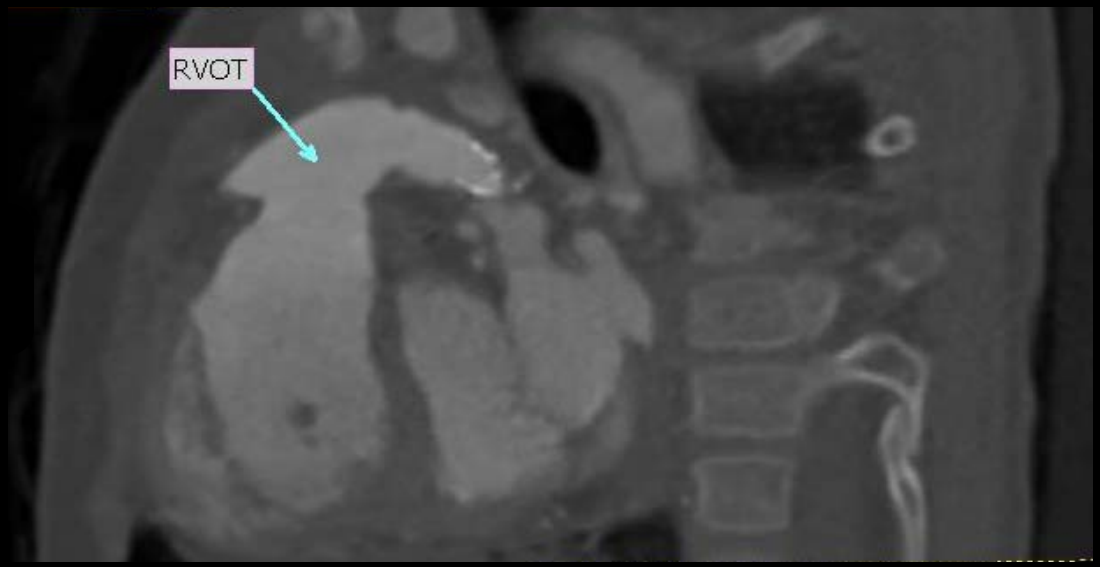
VSD: PMOE, large
← RV approach

Postop. pr. Ao (83/40/53)
RV (67/26/44)





Unifocalization, left



P(RV/LV)=0.8

Total repair

- 3yr LPA stent insertion
- 4yr RLPA, RUPA, LPA ballooning

CASE 3

- M / 6 mo
- PA,VSD, MAPCAs, No confluent central PA

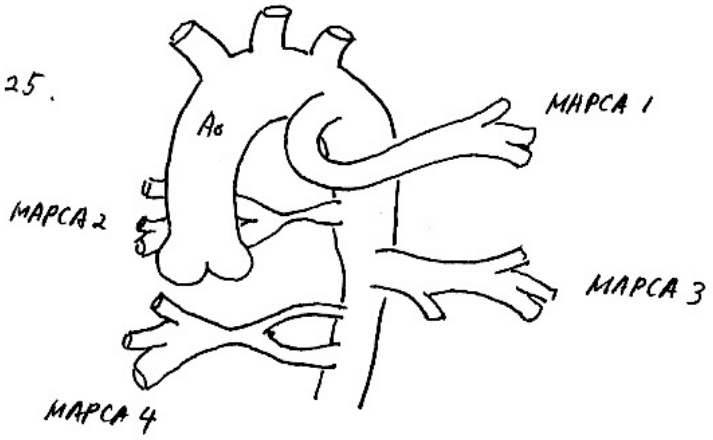
1st operation
6 mo/8.2 kg

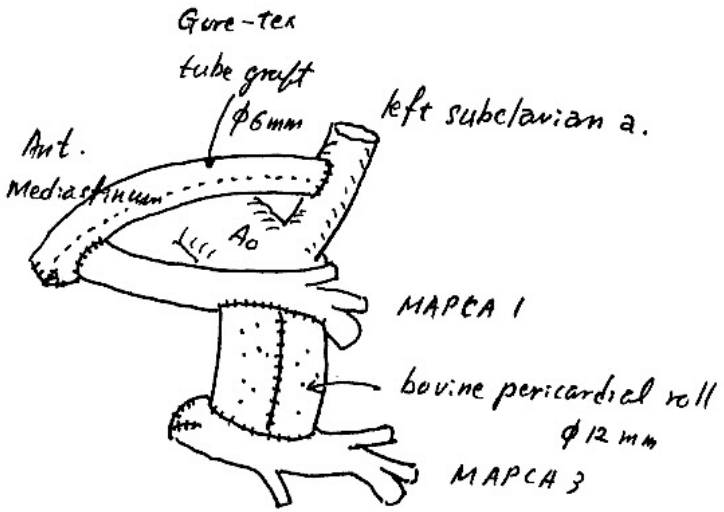
Left unifocalization
Left mod. BT shunt (6mm)

M/6 mo. 8.2kg)

PA, VSD, MAPCA (no confluent PA)

Op : 2007. 7. 25.





Left unifocalization

MAPCA 1. 3. : no protection, no collaterals (∵ no back flow)

MAPCA 2. 4 : stenosis (+)

2nd operation
13 mo/10.2 kg

Right Unifocalization
Right mod. BT shunt (5 mm)

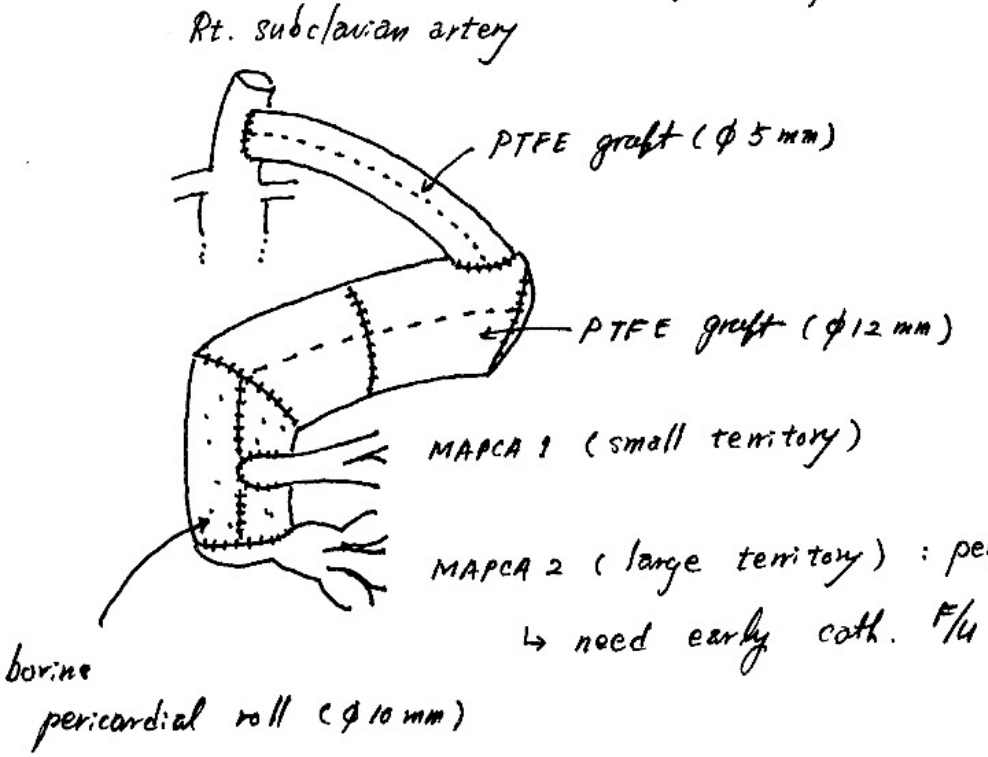
PA, VSD, MAPCA

s/p 2007. 7. 25. Left unifocalization

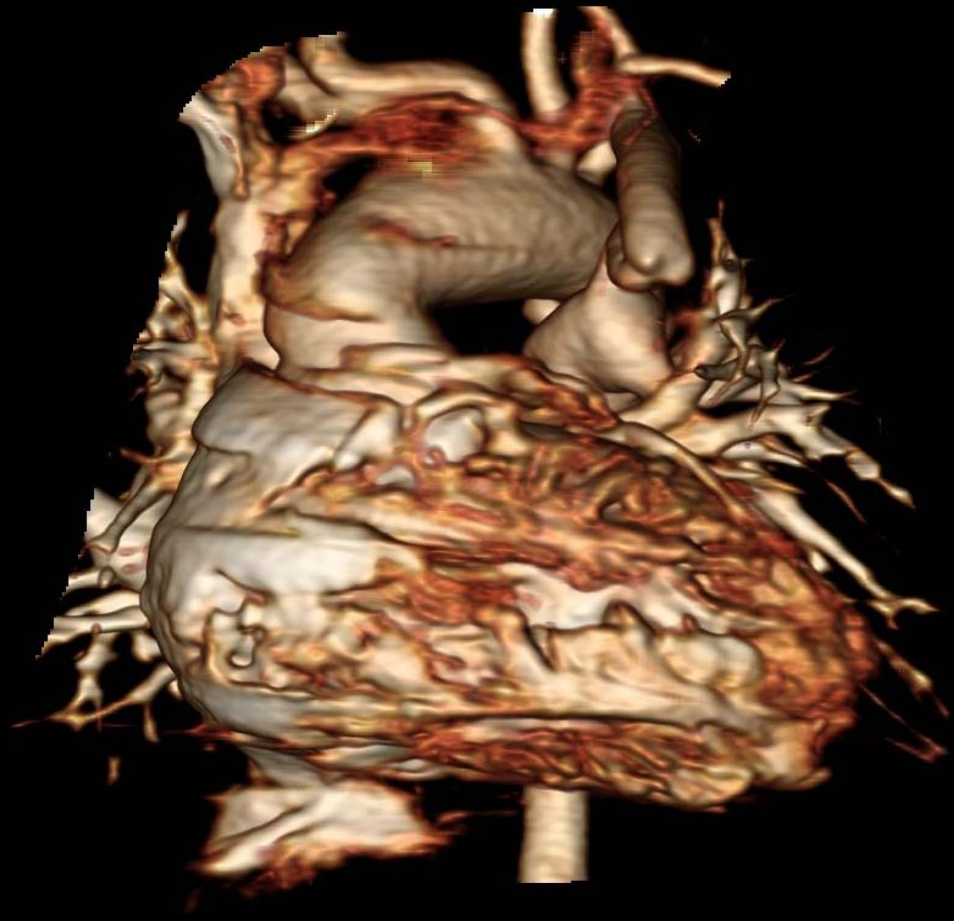
M/ 1yr. 6 mo.

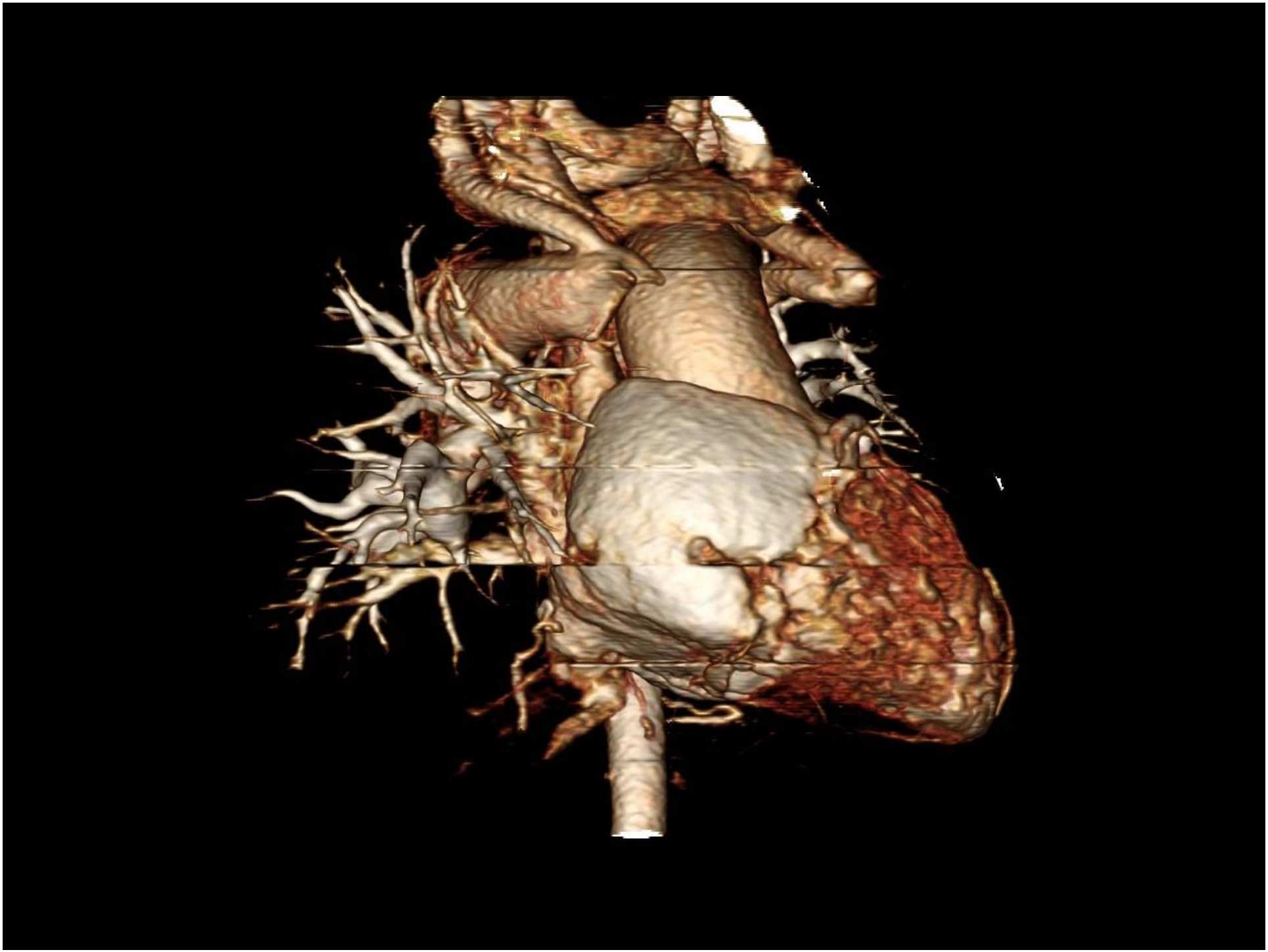
10.2kg

< Right unifocalization >



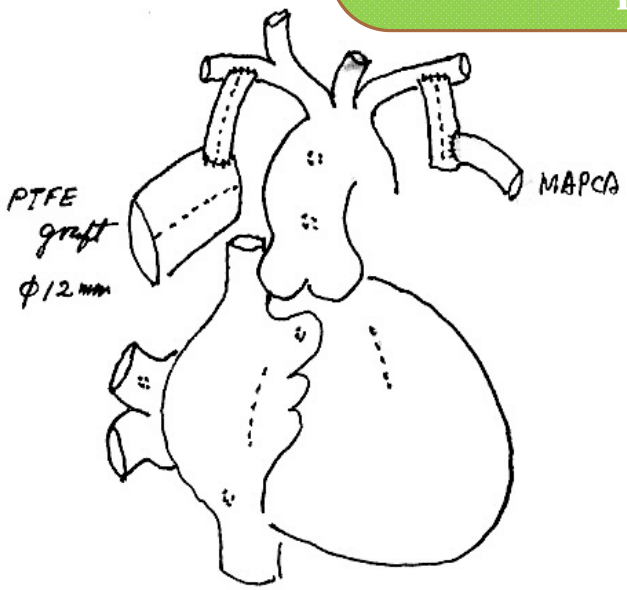
Lt. Rt. Unifocalization & BT shunt





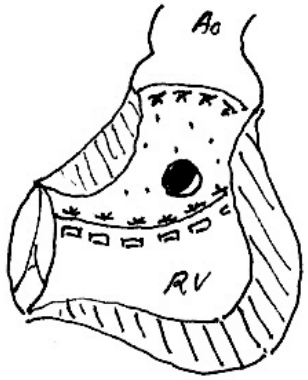
3rd operation
22 mo/10.6 kg

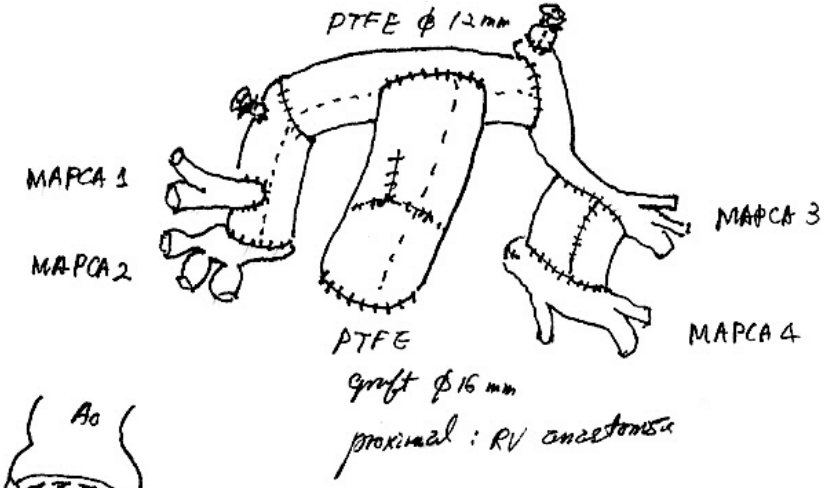
Rastelli op.
-Home-made PTFE valved conduit 16 mm
– Central confluent PA creation with PTFE tube graft 12 mm
– VSD patch baffling with fenestration (4mm)



PA, VSD, MAPCA (×4)

S/p 2007. 7. 25. left unifocalization
2008. 7. 28. right unifocalization

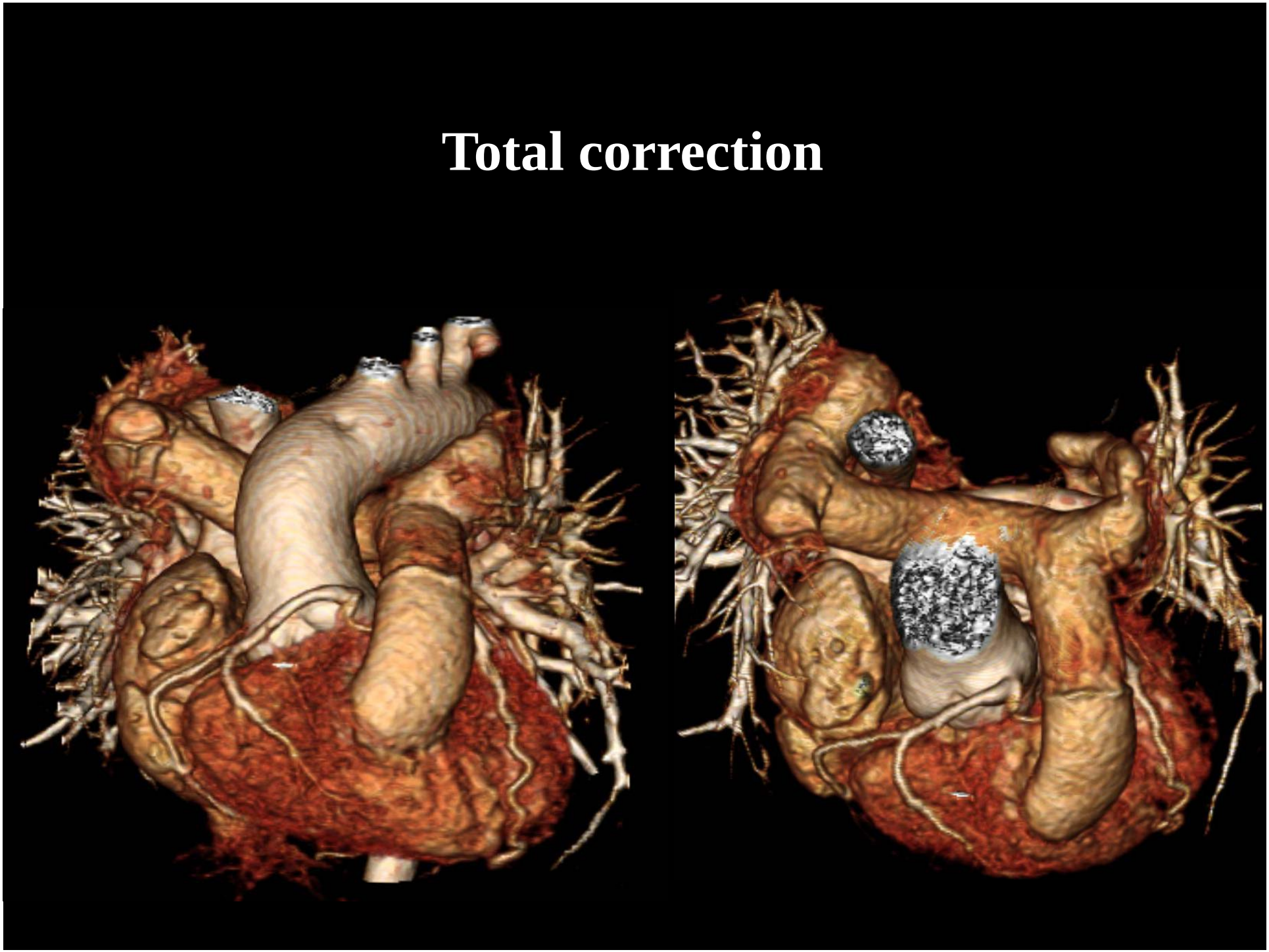




VSD : large PMVE

patch baffling via RV
→ fenestration φ4 mm

Total correction



CASE 4

- M/3 mo
- PA,VSD, MAPCAs
  - RPA : ascending aorta
  - LPA : descending aorta

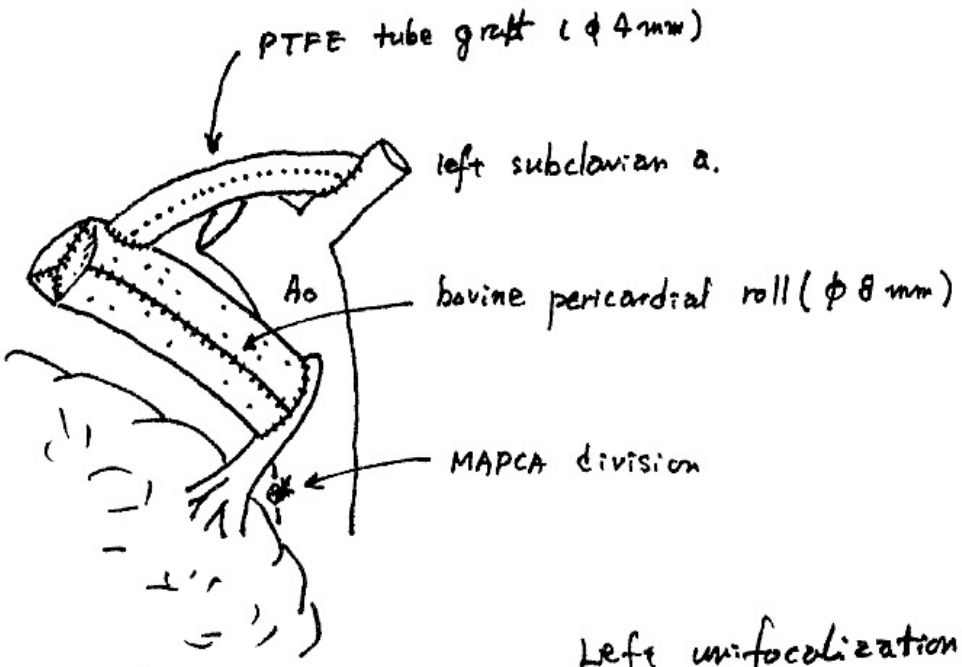
1st operation
3 mo/5.7 kg

Left unifocalization
- Bovine pericardial roll (8mm)
- Left mod. BT shunt (4mm)

op. : 2011. 1. 7.

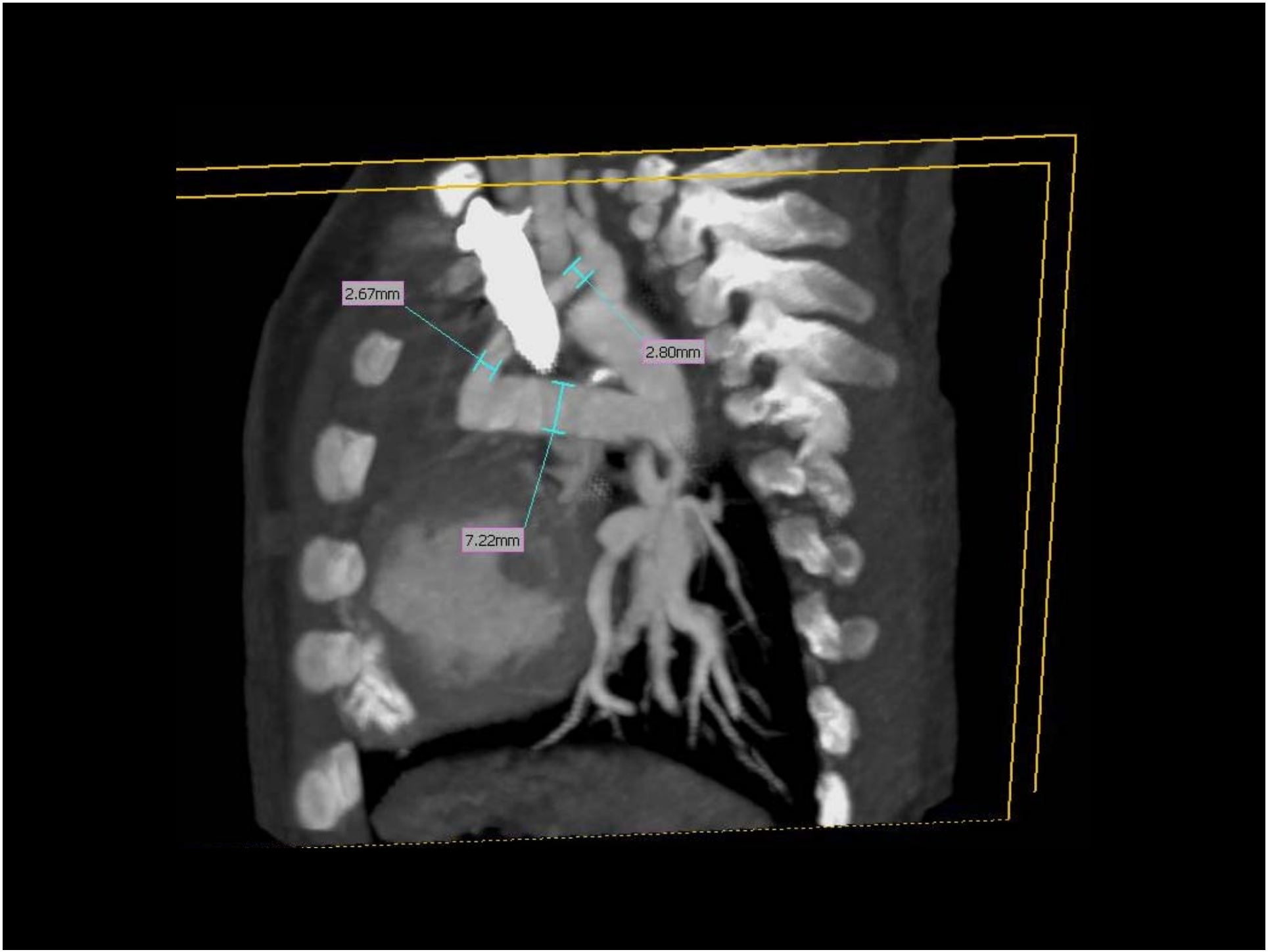
PA, VSD, MAPCA

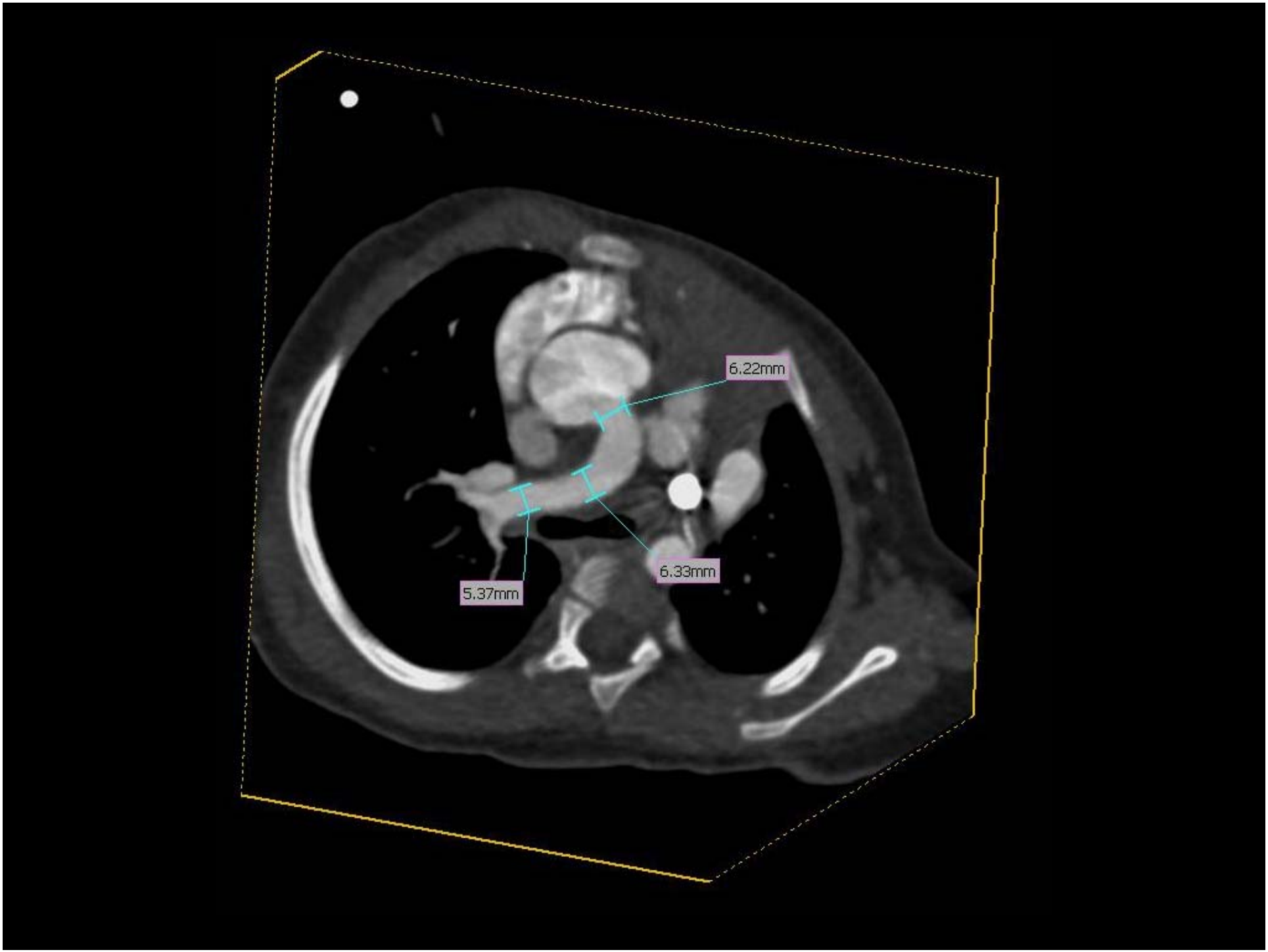
RPA from ascending aorta
LPA from descending aorta



LPA: hypoplastic, diffuse
ϕ 1.5 mm

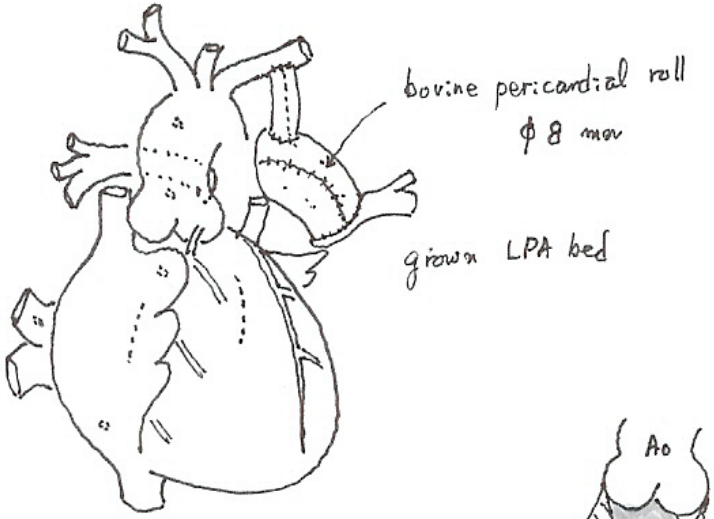
Left unifocalization


2.67mm
2.80mm
7.22mm


6.22mm
6.33mm
5.37mm

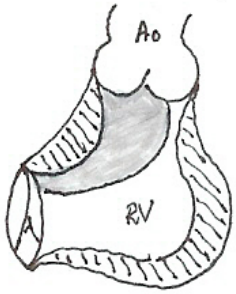
2nd operation
14 mo /8.5 kg

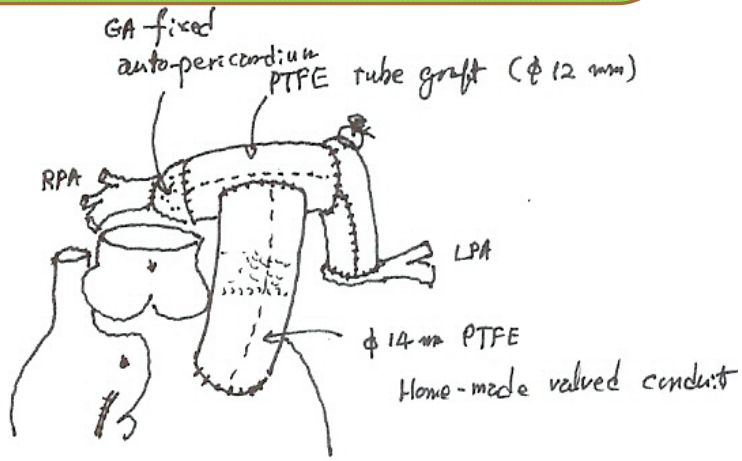
Rastelli op.
—VSD patch baffling
—RV-PA valved conduit interposition(Home-made PTFE valved graft 14 mm)
—Central PA reconstruction(PTFE tube graft 12mm, pericardial patch), Previous shunt division



PA, VSD, MAPCA
RPA from ascending aorta
LPA from descending thoracic aorta (φ1 mm)

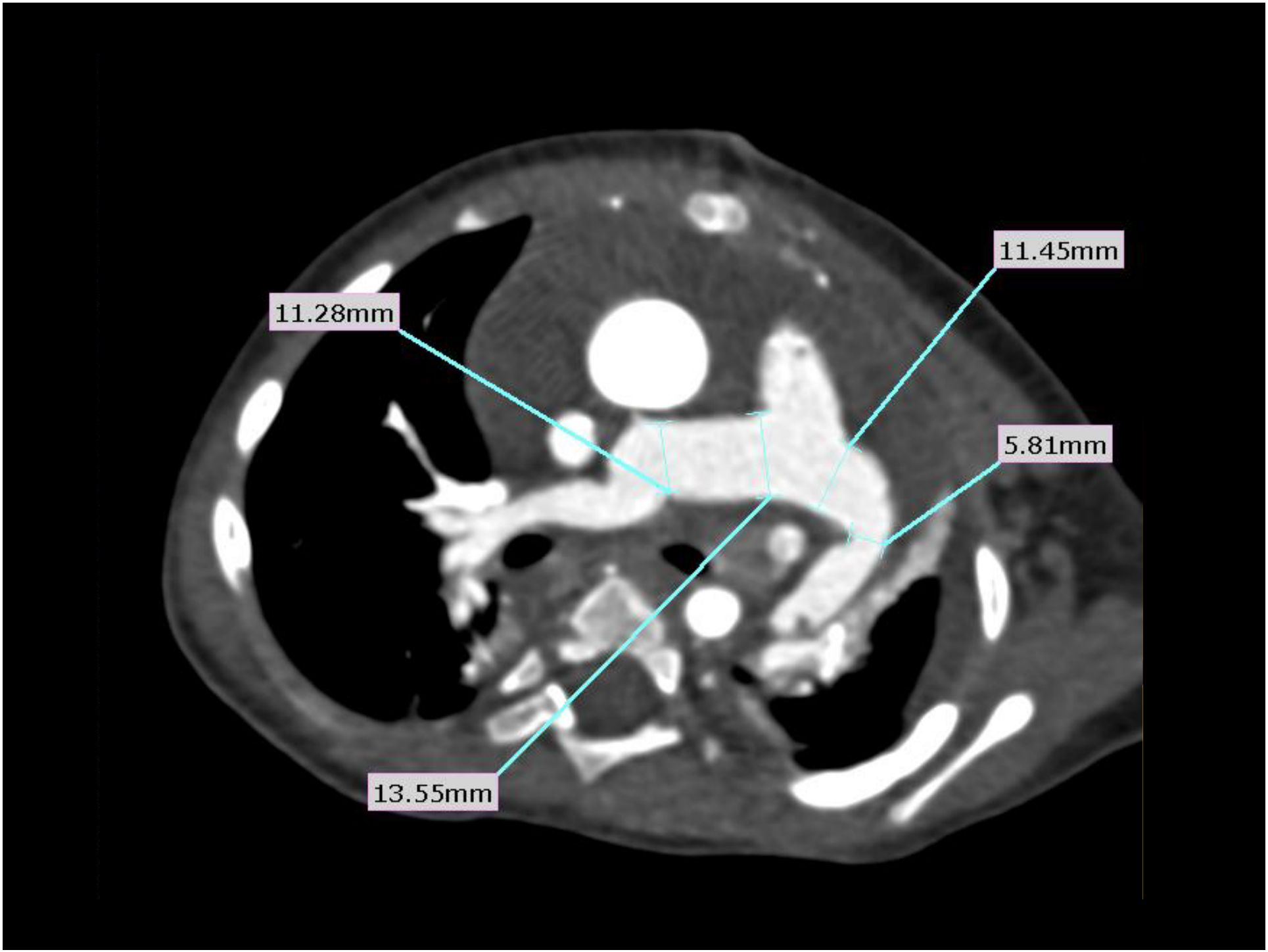
S/p 2011. 1. X. left unifocalization
left mod. B-T shunt (4 mm)





Rastelli op
1. VSD patch baffling via RV
2. RV-PA valved conduit interposition
Central confluent PA reconstruction
Previous B-T shunt division

Postop. $P_{RV/LV}$ = 0.7


11.45mm
11.28mm
5.81mm
13.55mm

CASE 5

- F/6 mo
- Rt. Isomerism

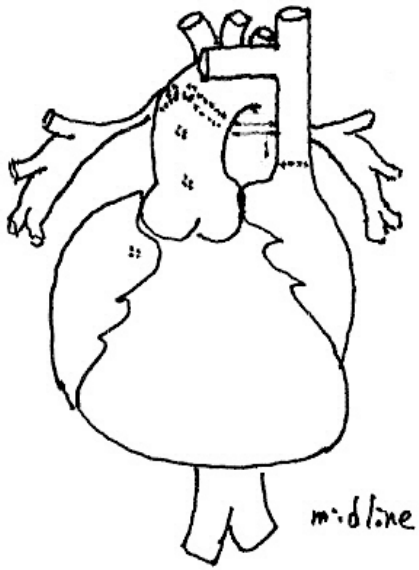
  UVH(RV type)

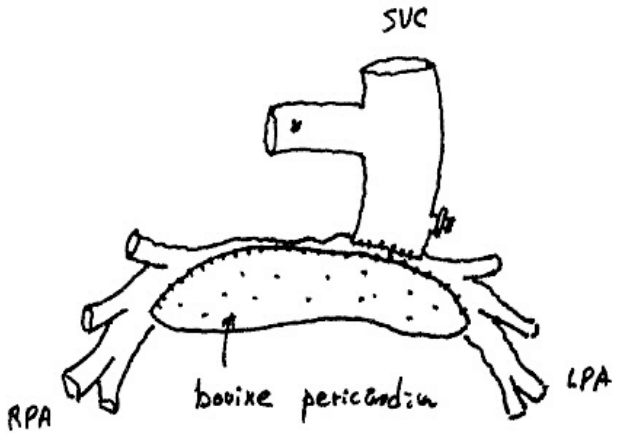
  MAPCA (x 2)

  No confluent PA
- 2011-04-12
  - BCPS, MAPCA unifocalization
  - Central confluent PA reconstruction (MAPCA+bovine pericardium)

F/6 mo./ 6.9 kg

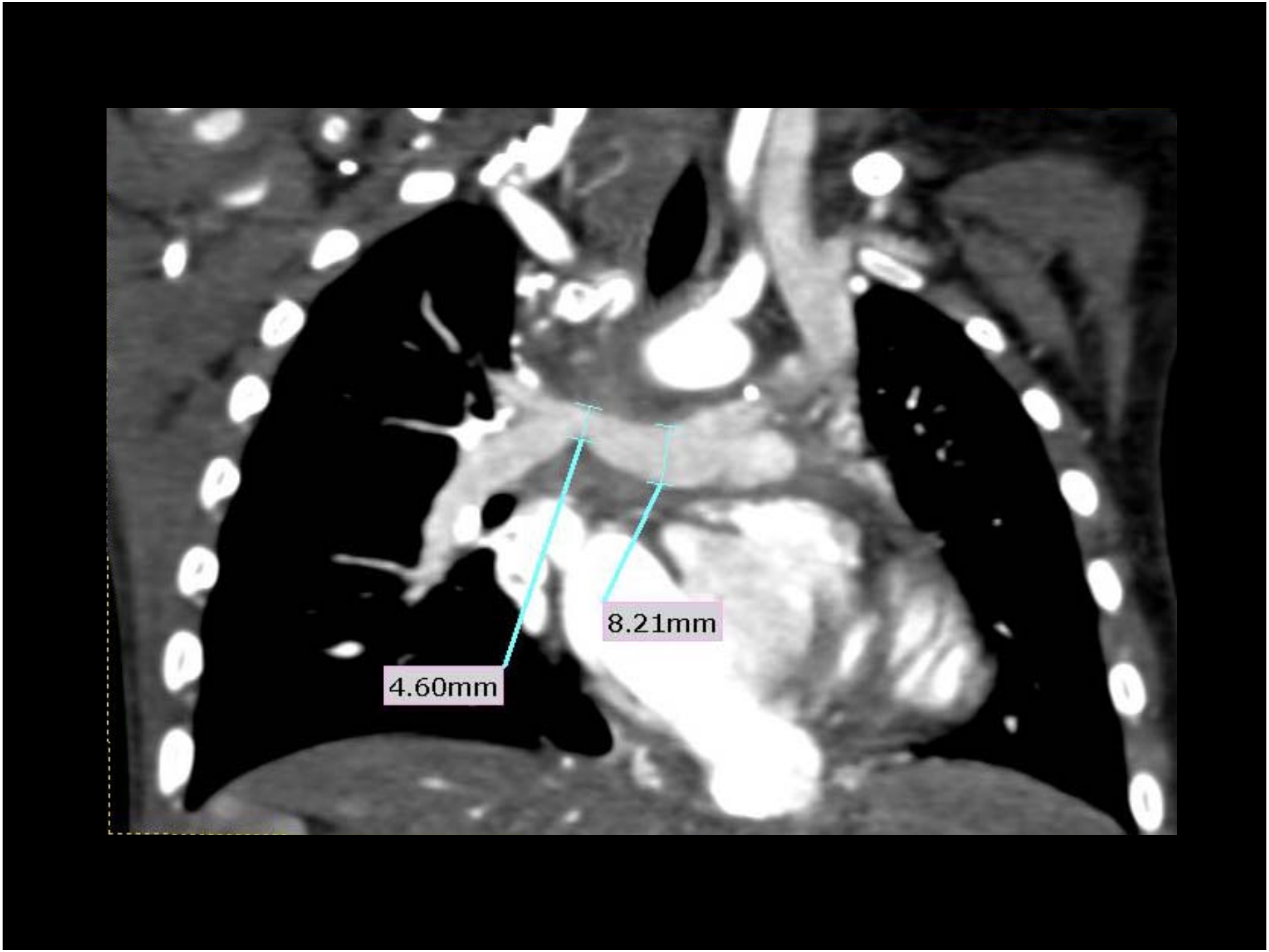
op. : 2011. 4. 12.





R/o Rt. isomerism
UVH (RV type). Pul. atresia
MAPCA (×2) : from proximal. aortic arch
No confluent central PA
Good hilum PA size

BCPS
MAPCA unifocalization
Central confluent PA reconstruction
(MAPCA + bovine pericardium)


8.21mm
4.60mm

Planned Surgery from Infant

- 11 patients (male=6, female=5)
- Mean FU duration
  - 45.9 ± 64.0 months
- Median age at initial operation
  - 6.0 months (Range 1.5-336.7 months)
- Median age at total correction
  - 1.9 yrs (Range 1.1-27.7 yrs)

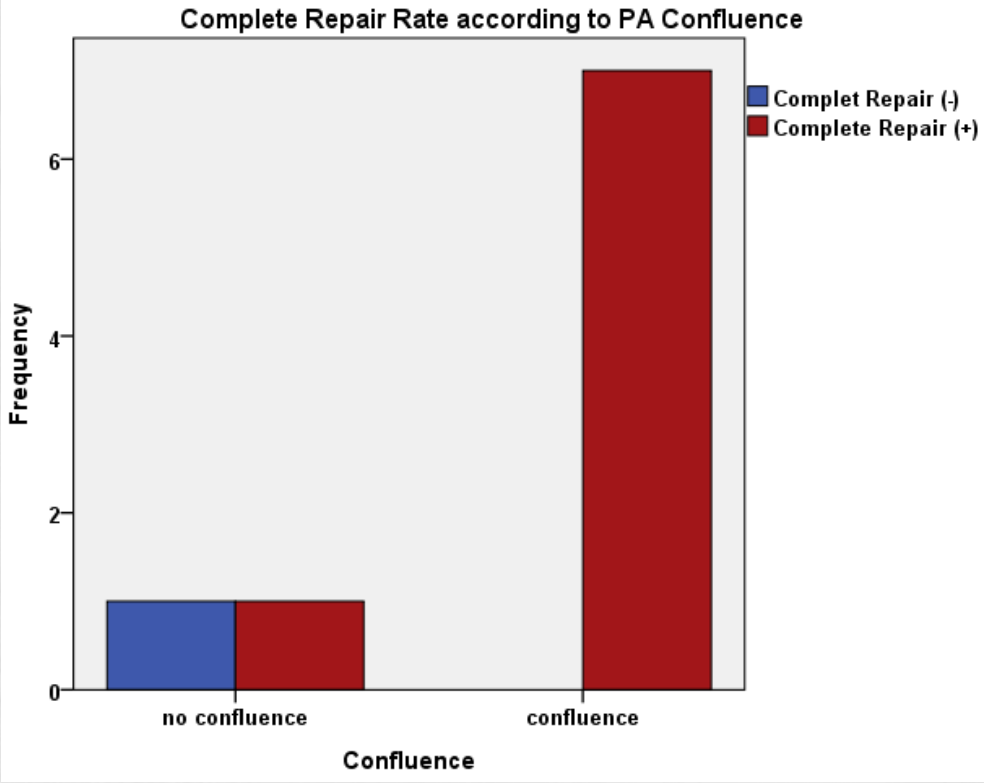
Planned Surgery from Infant

- MAPCA numbers/patient
  - Mean 3.5 ± 0.5
- Confluent central PA
  - 9 patients (9/11, 81.8%)

- Strategy
  - One stage total (n = 2) : success
  - Multi-stage approach (n = 9)
    - 1 mortality
    - 2 VSD fenestration
    - 2 waiting next state
- Overall complete repair rate (VSD closure)
  - 6 patients (6/9, 66.6 %)

Planned Surgery from Infant

- Complete Operation
  - Confluent PA
    - 100 % (7/7)
    - 1 VSD fenestration
  - Nonconfluent PA
    - 1 mortality
    - 1 VSD fenestration



Staged Reconstruction & Repair

Initial Palliation	No (%)
Shunt	1 (9.1%)
Central	1
BT shunt	0
RV-PA Conduit interposition	3 (27.3%)
RV-PA interposition	2
RV-PA interposition + Unifocalization	1
RV-PA interposition + Ligation	0
Unifocalization	5 (45.5%)
With BT	4
Without BT	1
Staged procedure number	11

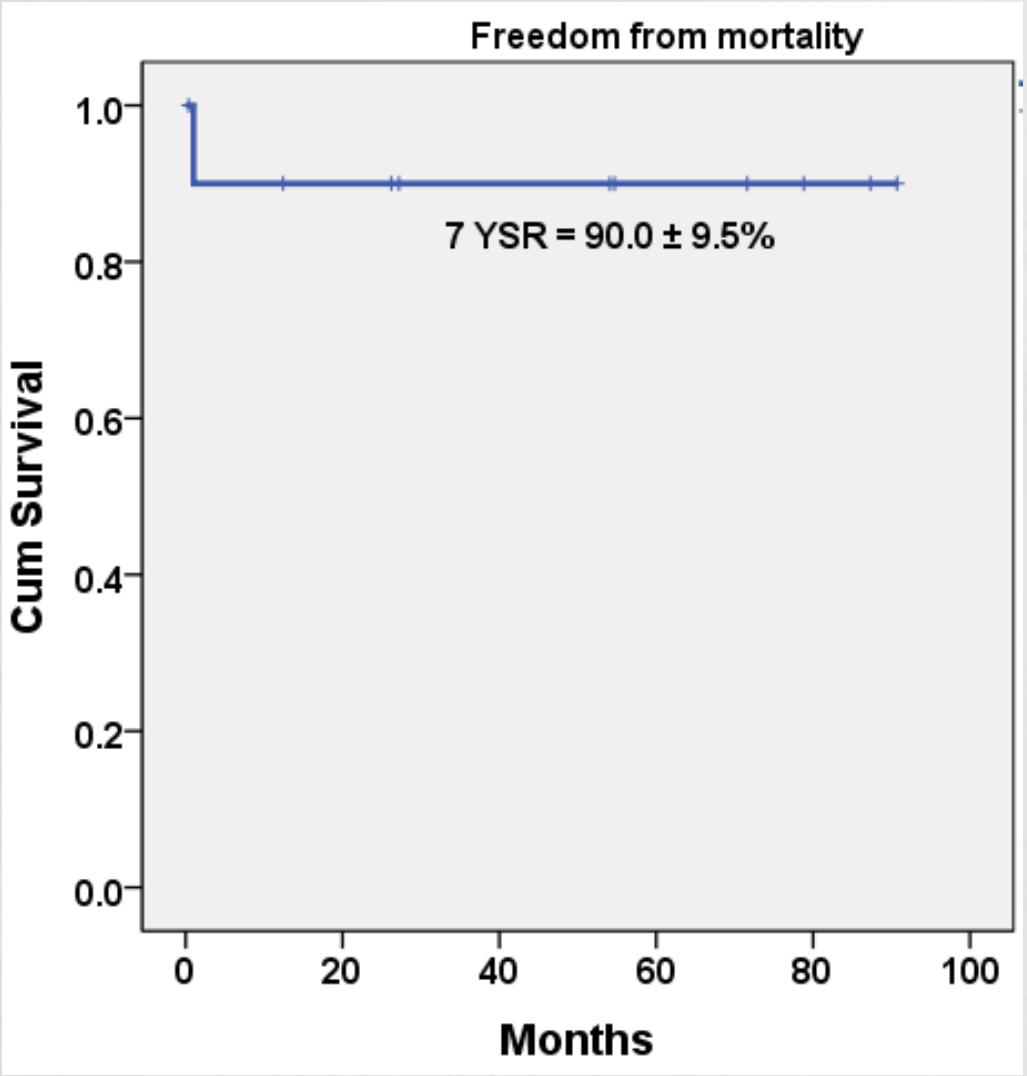
Management of MAPCAs

Manipulation of MAPCAs	No (%) (11 patients)
Unifocalization	6 (54.5%)
Ligation	2 (18.2%)
Unifocalization+ligation	2 (18.2%)
None	1 (9.1%)

Additional PA Procedures

Pulmonary artery procedures	Mean 1.5 ± 0.8
Surgical angioplasty	**8 patients**
Intervention	**2 patients**
Balloon angioplasty	4
Stent insertion	1

Planned Surgery from Infant



Conclusions

- One stage total correction may be successful in selected patients
- Confluent central pulmonary artery is most important factor for survival and complete repair.
- Individualized approach is required.