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
Melbourne Shunt

Central end-to-side Aortopulmonary shunt
Diminutive central pulmonary arteries
Modified Central Shunt
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
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 $\geq 50\%$ of predicted normal
  - *Puga JTCS 1989;98(6):1028-9*

- Predicted pRV/pLV $\leq 0.7$, No MAPCAs remain
  More than 2/3 lung segments are centralized
  - *by Iyer and Mee, ATS;1991:51:65-72*

- Nakata Index $> 150\text{mm}^2/\text{m}^2$ BSA
  - *by Metras, EJCTS 2001;20:590-6*

- TNPAI $\geq 200 \text{mm}^2/\text{m}^2$
  - *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 patients
    - CR (+) group
      - 1 patients - pneumonia
    - CR(-) group
      - Severe lung disease (CCAM)
#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

Mortality 1
- Persistent respiratory difficulty
- Prolonged ventilator care
- Desaturation and bradycardia
- 13mo, expired
Mortality 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**

17953126
F/27 / 57 kg

op. 12009.7.2. «Mid-line one-stage total repair»

MAPCA

TR (moderate)
AR (mild to moderate)
Ascending aorta dilatation

PA = VSD

Ascending aorta reduction-plasty

MAPCA 1
MAPCA 2
MAPCA 3
MAPCA 4

LCA

AV

TV

MAPCA 1
MAPCA 2
MAPCA 3
MAPCA 4

LPA

bovine pericardium

St. Jude Epic 25 mm
Inter Gard 26 mm

Rastelli op.

De Vega TAP

AV repair: Tidwell's method
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.
Melbourne shunt

1st operation
4 mo/5.4kg
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 (Ø1mm)

5/2006. 1. 12. MPA reimplantation to aorta

→ well growing PA

Palliative RV-PA conduit reconstruction (RV-PA conduit)
12month
- Ballon angioplasty of LPA and distal RPA
F / 1 yr. 8 mo.
3rd operation
21 mo/9.9 kg

Unifocalization of MAPCA, left

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

9/12 2006. 1. 12. MPA re-implantation 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
Total Repair
- Both PA patch angioplasty
- VSD patch baffling, RVOT patch widening
- RV infundibular muscle wide excision
- PFO primary closure
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)

M1 (6 mo. 8.2 kg)
PA, VSD, MAPCA (no confluent PA)

op: 2007. 7. 25.

MAPCA 1
MAPCA 2
MAPCA 3

MAPCA 1, 3: no protection, no collaterals (-: no back flow)
MAPCA 2, 4: stenosis (+)

Gore-tex
tube graft
φ5mm
left subclavian a.

Ant.
Mediastinum
L1, Ao

MAPCA 1
bovine pericardial roll
φ12mm
left unifocalization

MAPCA 3
2nd operation
13 mo/10.2 kg

Right Unifocalization
Right mod. BT shunt (5 mm)

M/1 yr. 6 mo.
10.2 kg

Rt. subclavian artery

PA, VSD, MAPCA

.dp 2007.7.25. Left unifocalization

< Right unifocalization >

PTEF graft (Φ 5 mm)

MAPCA 1 (small territory)

MAPCA 2 (large territory): peripheral PS (+)

 INSERT early cath. If necessary

pericardial roll (Φ 16 mm)
Lt. Rt. Unifocalization & BT shunt
Rastelli op.
- Home-made PTFE valved conduit 16 mm
  - Central confluent PA creation with PTFE tube graft 12 mm
  - VSD patch baffling with fenestration (4 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. X.

PA, VSD, MAPCA
RPA fr. ascending aorta
LPA fr. descending aorta

PTFE tube graft (ø 4mm)
left subclavian a.

Ao
bovine pericardial roll (ø 8 mm)

MAPCA division

LPA, hypoplastic, diffuse
ø 1.5 mm

Left unifocalization
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 12 mm, pericardial patch), Previous shunt division
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.

midline IVC & HV

BCPS
MAPCA uni-focalizatjon
Central confluent PA reconstruction
(MAPCA + bovine pericardium)

Rv. Re. isomerism
UVH (RV type), Pul. atresia
MAPCA (x2): from proximal aortic arch
No confluent central PA
Good hilar PA size
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 $3.5 \pm 0.5$
- 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

<table>
<thead>
<tr>
<th>Initial Palliation</th>
<th>No (%)</th>
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<tbody>
<tr>
<td><strong>Shunt</strong></td>
<td></td>
</tr>
<tr>
<td>Central</td>
<td>1 (9.1%)</td>
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<tr>
<td>BT shunt</td>
<td>0</td>
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<tr>
<td><strong>RV-PA Conduit interposition</strong></td>
<td></td>
</tr>
<tr>
<td>RV-PA interposition</td>
<td>3 (27.3%)</td>
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<tr>
<td>RV-PA interposition + Unifocalization</td>
<td>2</td>
</tr>
<tr>
<td>RV-PA interposition + Ligation</td>
<td>1</td>
</tr>
<tr>
<td><strong>Unifocalization</strong></td>
<td></td>
</tr>
<tr>
<td>With BT</td>
<td>4</td>
</tr>
<tr>
<td>Without BT</td>
<td>1</td>
</tr>
<tr>
<td><strong>Staged procedure number</strong></td>
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<tr>
<td></td>
<td>11</td>
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## Management of MAPCAs

<table>
<thead>
<tr>
<th>Manipulation of MAPCAs</th>
<th>No (%) (11 patients)</th>
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<tbody>
<tr>
<td>Unifocalization</td>
<td>6 (54.5%)</td>
</tr>
<tr>
<td>Ligation</td>
<td>2 (18.2%)</td>
</tr>
<tr>
<td>Unifocalization+ligation</td>
<td>2 (18.2%)</td>
</tr>
<tr>
<td>None</td>
<td>1 (9.1%)</td>
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# Additional PA Procedures

<table>
<thead>
<tr>
<th>Pulmonary artery procedures</th>
<th>Mean 1.5 ± 0.8</th>
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<tbody>
<tr>
<td>Surgical angioplasty</td>
<td>8 patients</td>
</tr>
<tr>
<td>Intervention</td>
<td>2 patients</td>
</tr>
<tr>
<td>Balloon angioplasty</td>
<td>4</td>
</tr>
<tr>
<td>Stent insertion</td>
<td>1</td>
</tr>
</tbody>
</table>
Planned Surgery from Infant

Freedom from mortality

7 YSR = 90.0 ± 9.5%
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.