PA c IVS
THE EVALUATION AND PRINCIPLES
OF TREATMENT STRATEGY
PA c IVS (not only pulmonary valve disease)

Pathophysiology

- Atretic pulmonary valve with diaphragmatic membrane (80%)
- Infundibular atresia (20%)
- Annulus and MPA – hypoplastic or normal
- RV size varies and related to survival
- Coronary artery anomaly (ventriculocoronary connection, coronary sinusoid, proximal coronary artery obstruction)
- Interatrial communication, PDA - essential
Evaluation

- Echocardiography
- Cardiac catheterization
Echocardiography

- Atretic pulmonic valve – Doppler evidence
- RV hypertrophy, small cavity
- TV size measure
- ASD (or Rt to Lt shunt PFO), PDA
- Branch PA
- Coronary circulation
Echocardiography

- Integral tool for the diagnosis – fetus and neonate
- Segmental analysis
- Systemic, pulmonary venous return (bilateral SVC, interruption of IVC with azygous communication – important Glenn, Fontan)
- Atrial communication – Rt to Lt, obstruction (5-10%)
- TV morph. size – RV size, coronary anomaly, RV growth (correlation)

Drant SE. Pediatr Cardiol 2001
Echocardiography (TV evaluation)

- Hypoplasia of valve annulus
- Shortening and thickening of the chordae
- Myxoid change of the leaflets

Drant SE. Pediatr Cardiol 2001
Echocardiography (TV evaluation)

- TV annulus \textit{Z-score}; closely correlated to RV size and RV to coronary communication
- Doppler study
  - tricuspid inflow signals – with decreasing of the color scale
  - tricuspid insufficiency jet velocity – estimate the RV pressure

\textit{Hanley FL et al. J Thorac Cardiovasc Surg 1993}
\textit{Drant SE. Pediatr Cardiol 2001}
Standard score (Z-score)

Indicates how many standard deviations an observation is above or below the mean
TV Z-score

Daubney et al. JACC 2002

Kirklin JW. Cardiac Surgery. 2nd ed. 1993 p30
Tricuspid valve size

- TV Z-score $\geq -2.5$ for BSA, tricuspid/mitral valve ratio $\geq 0.7$; no risk of RV dependent coronary circulation, achieving biventricular repair
- Highly associated with survival rate among the biventricular repair group
- $Z$-score $> -2.4$ for BSA; biventricular repair

De Leval M et al. Circulation 1985
Echocardiography (RV)

- 3 portions of RV (inlet, trabecular, infundibulum)
- 3 types; Tripartite, Bipartite, monopartite
- RV size – highly correlated with TV size
RV size and morphology

- Quantitative assessment; RV volume in subcostal coronal and sagittal view (by biplane Simpson’s rule)
- Qualitative assessment; RV inflow portion – compare with LV (apical view)
- The patency and size of RV outflow tract; important for RV to coronary communication and successful decompression

*Trowitzsh E et al. J Am Coll Cardiol 1985*
*Billingsly AM et al. J Torac Cardiovasc Surg 1989*
*Mainwaring RD et al. J Thorac Surg 1993*
Pulmonary valve, MPA, br.PA

- Parasternal views
- Atretic pulmonary valve, pinhole patency
- MPA, branches – usually well developed, retrograde flow via ductus arteriosus
- Absence of PA – AP collaterals, MAPCA
- PDA flow – initial decision-making of PGE₁
Coronary circulation (RV communication)

Emmel M et al. Heart 2004;90:94
Coronary circulation (RV communication)

- Most important determinants of early management
- RCA, LAD – parasternal long, short axis
- Coronary angiography – remains necessary to completely evaluation
- TV annulus size (Z-score) and morphology of RV outflow tract (infundibulum) – strong correlation

Satou GM et al. Am J Cardiol 2000
Drant SE. Pediatr Cardiol 2001
Coronary circulation (pathology)

- RV – coronary sinusoid (60-70%)
- RV – coronary connection (45-55%)
- Coronary stenosis or interruption (10%)
- Absence of proximal aorto-coronary connection (15%)
- One coronary artery from PA
RV dependent coronary circulation

- intrinsic coronary lesion (stenosis, interruption) – proximal area
- the distal coronary flow – completely RV dependent
- RV decompression – lead to ischemia, infarction and death
- associated with diminutive and hypertensive ventricle, negative TV Z-score, infundibular atresia, LVOTO and LV dysfunction on follow up

Dyamenahalli U et al. Cardio Young 2004
박인숙. 선천성 심장병 2nd ed. p 547
Left side heart

- Mitral valve, LV wall motion – apical, parasternal view
- Suprasystemic RV pressure, RV dependant coronary circulation; LV wall motion abnormality, myocardial ischemia
- LV dysfunction, MV or AV anomaly – rare
Echocardiographic Predictors of Outcome in Fetuses with Pulmonary Atresia with Intact Ventricular Septum

Renuka E. Peterson, MD, Daniel S. Levi, MD, Ryan J. Williams, Wyman W. Lai, MD, MPH, Mark S. Sklansky, MD, and Stacey Drant, MD, Los Angeles, California, and New York, New York

Objective: We sought to identify in utero predictors of postnatal outcomes in fetal patients with pulmonary atresia with intact ventricular septum (PAIVS) or critical pulmonary stenosis.

Background: Although PAIVS or critical pulmonary stenosis can be diagnosed in utero by echocardiography, our ability to predict outcomes is limited.

Methods: Fetal echocardiograms from 28 patients with PAIVS/critical pulmonary stenosis were retrospectively reviewed. Tricuspid valve (TV) annulus, right and left ventricular internal dimensions, and degree of tricuspid regurgitation were recorded. To establish normal fetal values, echocardiograms from healthy patients were analyzed in an identical fashion.

Results: Both a fetal TV - score of -4 or less beyond 23 weeks of gestation and a fetal TV annulus of 5 mm or less beyond 30 weeks of gestation were predictive of poor postnatal outcomes. In addition, right-left ventricular length or width less than 0.5 and/or the absence of tricuspid regurgitation were predictive of poor outcome.

Conclusions: TV annulus size, right-left ventricular ratios, and presence of tricuspid regurgitation on fetal echocardiograms may aid in guiding prenatal counseling regarding postnatal outcome in PAIVS.

([J Am Soc Echocardiogr 2006;19:1393-1400.])

Children’s Hospital of LA, UCLA
Determinants of Outcome in Fetal Pulmonary Valve Stenosis or Atresia With Intact Ventricular Septum

Kevin S. Roman, MD\textsuperscript{a}, Jean-Claude Fouron, MD\textsuperscript{b}, Masaki Nii, MD\textsuperscript{a}, Jeffrey F. Smallhorn, MBBS\textsuperscript{a}, Rajiv Chaturvedi, MD\textsuperscript{a}, and Edgar T. Jaeggi, MD\textsuperscript{a,*}

Pulmonary valve stenosis or atresia with intact ventricular septum represents a spectrum of severity. This study aimed to identify ultrasound markers of biventricular versus non-biventricular outcome. The fetal echocardiograms of 41 fetuses diagnosed with pulmonary stenosis or atresia and right ventricular (RV)/left ventricular (LV) length ratios $>0.4$ from 17 to 31 weeks of gestation were reviewed. Of 27 live-born patients with intention to treat, 8 had non-biventricular outcomes and 19 had biventricular circulation. At the time of diagnosis, poor RV function, flow reversal in the arterial duct, the degree of tricuspid valve (TV) regurgitation, and inferior vena cava Doppler flow pattern did not differ between the 2 outcome groups. However, RV sinusoids, the RV/LV length ratio, the TV/mitral valve ratio, and TV inflow duration were significantly different. Cut-off values derived from receiver-operating characteristic curves yielding the best sensitivity and specificity for a non-biventricular outcome were TV/mitral valve ratio $<0.7$, RV/LV length ratio $<0.6$, TV inflow duration $<31.5\%$ of cardiac cycle length, and the presence of RV sinusoids. If 3 of these 4 criteria were fulfilled, this predicted a non-biventricular outcome with sensitivity of 100\% and specificity of 75\%. In conclusion, in fetuses $\leq 31$ weeks of gestation with pulmonary stenosis or atresia and intact ventricular septum, progression to a non-biventricular outcome can be predicted by a 4-criterion scoring system. The criteria may be useful in selecting fetuses for prenatal catheter intervention to prevent progressive RV hypoplasia. © 2007 Elsevier Inc. All rights reserved (Am J Cardiol 2007;99:699–703)
Catheterization

- Hemodynamics – RV, RA pressure and O2 sat
- RV angiogram – RV morphology, size, coronary fistula
- Aortography – arch, PDA, MAPCA, coronary angio
- Balloon valvotomy – potential opening, check RV dependent coronary circulation
Treatment strategies

- Goal of treatment
  - separate pulmonary and systemic circulation
  - eliminate cyanosis
  - without compromising cardiac output
  - without inducing systemic venous hypertension

→ Achieve by univentricular, biventricular, one and half ventricular repair

Bichell DP. Curr Opin Cardiol 1999
Treatment Algorithm

Newborn with P Atresia with IVS

PGE1

No Ebstein
No RVDDCC

Balloon Dil of P valve or RVOTR/Valvotomy ± Shunt
No BAS

Echo @ age 2-3 mo.

If residual RVOTO(+)

Balloon Dil or RVOTR

No Residual RVOTO
(wide open RVOT)

"Good" RV/TV

"Too small" RV/TV

BCS ≥ age 5-6 mo

Small or no ASD

Test Occlusion of ASD

If OK

AsD Closure ± Shunt Closure (BVR)

No More Procedure ± Shunt Closure

If Fail

1 & 1/2 V Repair (BCS, RVOTR, AsD Closure)

Fontan ≥ age 2 yrs

Ebstein or RVDDCC or very small RV/TV

Shunt & BAS for small or no ASD

박인숙. 선천성 심장병 2nd ed. p 556
# Morphologic classification and treatment strategies

## Group A
- **Good RV anatomy**
  - TV z score >-2.5
- Usually tripartite RV
- Membranous atria (good infundibulum)
- Usually no major sinusoids
- Variable tricuspid regurgitation

## Group B
- **Intermediate anatomy**
  - Borderline RV size
    - (TV z score -2.5 to -4.5)
  - Usually bipartite RV – absent or markedly attenuated trabecular component
  - Patent infundibulum
  - Major sinusoids may be present
  - Minor sinusoids common
  - Small PV annulus
  - Subvalvar stenosis
  - Variable tricuspid regurgitation

## Group C
- **Severe RV hypoplasia**
  - TV z score <-5.0
  - Unipartite – absent trabecular component and diminutive/absent infundibulum
    - (muscular atresia)
  - Hypoplastic TV
  - Usually competent TV
  - Major sinusoids more common
  - Stenoses or interruption

### Treatment at presentation
- **Group A**
  - RFV (primary procedure)

- **Group B**
  - RFV + PDA stenting
  - ± Balloon atrial septostomy (primary procedure)

- **Group C**
  - PDA stenting and balloon atrial septostomy or systemic-pulmonary shunt

### Reinterventions uncommon
- Repeat balloon dilation if restenosis occurs
- RVOT reconstruction if progressive fixed stenosis occurs
- Tricuspid valve repair if progressive tricuspid regurgitation

### Surgical re-interventions
- **Group B**
  - Bidirectional Glenn shunt (1½ ventricle) if RV growth is poor
  - RVOT reconstruction for subvalvar obstruction/smal PV annulus
  - TV repair for severe or progressive tricuspid regurgitation

- **Group C**
  - No cyanosis: no further intervention
  - Cyanosis: device closure of PFO/ASD

### Good/fair RV growth
- Surgery (primary procedure)
  - Bidirectional Glenn shunt
  - Fontan completion

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*Alwi M. Catheter Cardiovasc Interv. 2006*
Post procedural evaluation

- Systemic to pulmonary shunt
- Post RV decompression
Systemic to pulmonary shunt

- Without RV decompression
- RV dependent coronary circulation
- Follow up focus – patency of shunt and LV function
- Intimal proliferation at the sites of RV to coronary fistula – LV dysfunction
- RV, TV – do not grow (relative small)
Post RV decompression

- Early evaluation assessing need for additional shunt
- Adequacy of decompression and RV pressure
- Assessment of growth of the RV and TV
- Assessment of LV function

Drant SE. Pediatr Cardiol 2001
Assessing need for additional shunt

- Early post op period – elevated pulmonary vascular resistance and poor RV compliance
  - limit pulmonary blood flow
- Prolonged use of PGE1 or additional shunt
- Echo predictors – RV end diastolic volume, TV annulus diameter

Trowitzsch E et al. J Am Coll Cardiol 1985
Adequacy of decompression and RV pressure

- Establishing patency of RV outflow tract and pulmonary valve – 2D imaging from subcostal parasternal views
- Dynamic narrowing with systole (acute reduce of afterload)
- Quantified measurement of RVOT and pulmonary valve – color Doppler (peak and mean pressure gradient)
- Measuring the peak gradient of TR jet
Assessment of growth of the RV and TV

- Check the early change of RVEDV, RV area ratio, stroke volume
- RV cross sectional area obtained from the ventricular length and width – apical, subcostal 4 ch. View
- The change of TV annulus size

Schmidt KG et al. J Am Coll Cardiol 1992
Hanseus K et al. Pediatr Cardiol 1991
Assessment of LV function

- RV dependent coronary circulation
- Progressive intimal hyperplasia within the coronary artery
  → early LV dysfunction after RV decompression
- Unsuccessful RV decompression
Long term follow up evaluation

Late Pulmonary Valve Replacement in Patients With Pulmonary Atresia and Intact Ventricular Septum: A Case-Matched Study

Victor Bautista-Hernandez, MD, PhD,* Babar S. Hasan, MD,* David M. Harrild, MD, Ashwin Prakash, MD, Diego Porras, MD, John E. Mayer, Jr, MD, Pedro J. del Nido, MD, and Frank A. Pigula, MD

Departments of Cardiac Surgery and Cardiology, Children’s Hospital Boston, and Departments of Surgery and Pediatrics, Harvard Medical School, Boston, Massachusetts

Background. Pulmonary valve replacement (PVR) is a common therapy for chronic pulmonary regurgitation. However, the use of this strategy is mostly based on the studies performed on patients with tetralogy of Fallot (TOF) and not in patients with pulmonary atresia/intact ventricular septum (PA/IVS). The aim of this study is to evaluate our experience with PVR in patients with PA/IVS and compare them with a matched cohort of TOF patients.

Methods. Between 1995 and 2009, 13 patients with PA/IVS underwent a late PVR. Matched TOF control subjects were identified for 12 of these patients. Before and after PVR, echocardiographic, magnetic resonance imaging, exercise test, Holter, and electrocardiographic data were compared between groups.

Results. There was no mortality in either group. The PVR improved pulmonary regurgitant fraction and right ventricular volumes in all patients. Patients with PA/IVS had more significant tricuspid regurgitation (TR [at least moderate]) by echocardiography and magnetic resonance imaging before PVR (n = 11 [85%] versus n = 1 [8%]; p = 0.003) and had more tricuspid valve repairs than TOF patients (n = 9 [69%] versus n = 1 [8%]; p = 0.004). Repair was undertaken by a combination of techniques. Although TR was improved early postoperatively, only 2 of 9 patients (22%) were free from significant TR at most recent follow-up (median 2.5 years; range, 0.1 to 10.9). No patient underwent reoperation at latest follow-up.

Conclusions. Patients with PA/IVS can undergo a late PVR with excellent results. Significant TR and repair are more commonly observed among patients with PA/IVS compared with TOF patients. Although tricuspid valve repair improves regurgitation early, TR tends to recur, suggesting the need for further refinement of current surgical techniques.

Clinical outcomes of adult survivors of pulmonary atresia with intact ventricular septum

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b Division of Cardiology, Children’s National Medical Center, George Washington University, Washington, DC, United States

<table>
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<th>Characteristics</th>
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<th>Table 1: Demographics and initial operative strategies of PA/IVS.</th>
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<th>Univentricular, Fontan (n=7)</th>
<th>Biventricular repair (n=8)</th>
<th>Total number (n=20)</th>
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<td>Average age, years</td>
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<td>Left ventricular EF</td>
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<td>51% (45–58%)</td>
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Long term follow up evaluation for PA c IVS | echocardiography, EKG, Holter, MRI

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