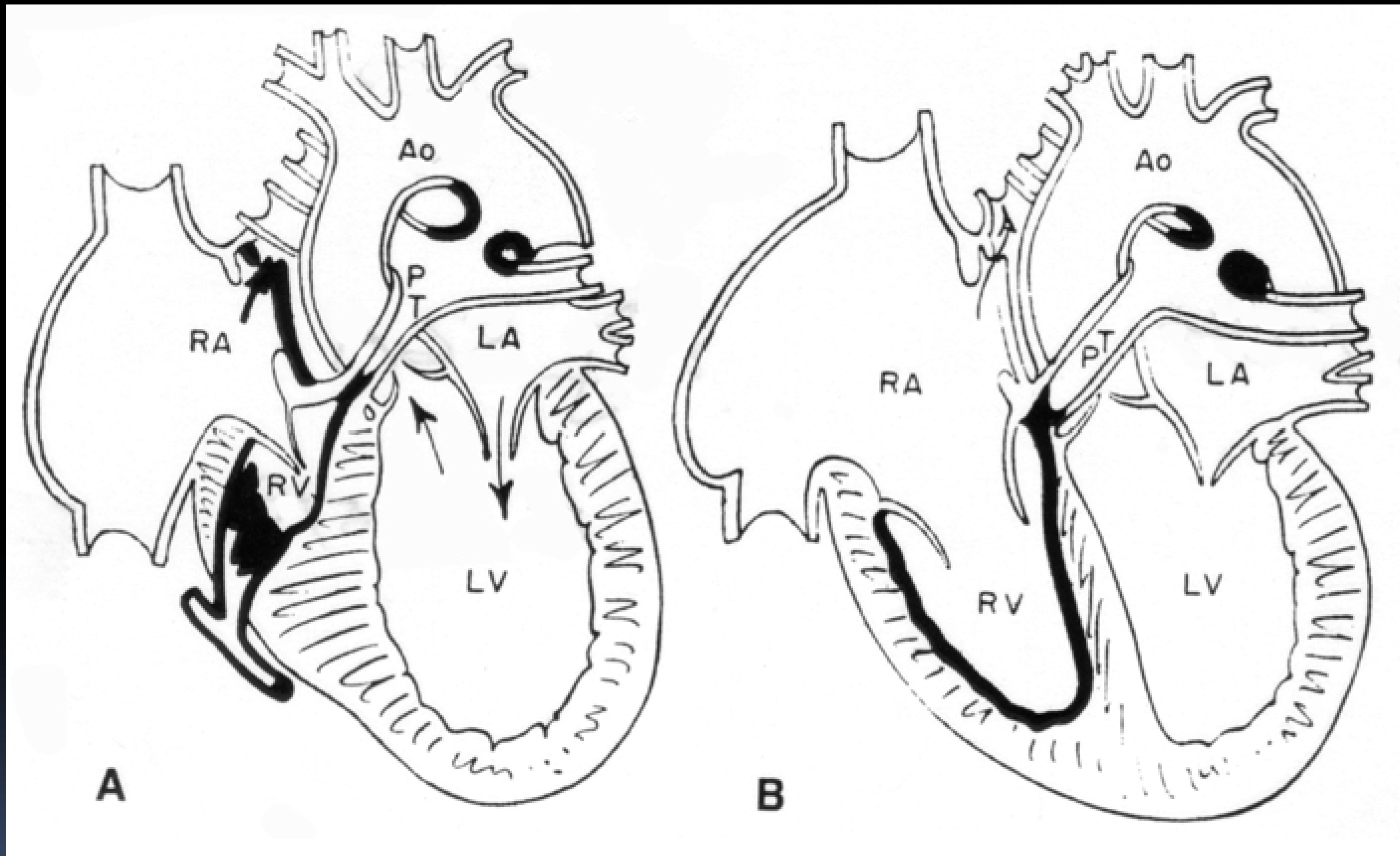
가천의대 길병원 소아심장과 최덕영

PA c IVS
THE EVALUATION AND PRINCIPLES OF TREATMENT STRATEGY

PA c IVS (not only pulmonary valve disease)



Edwards JE. Pathologic Alteration of the right heart. In: Konstam MA, Isner M, eds. The right ventricle. The Hague: Martinus-Nijhoff, 1987

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

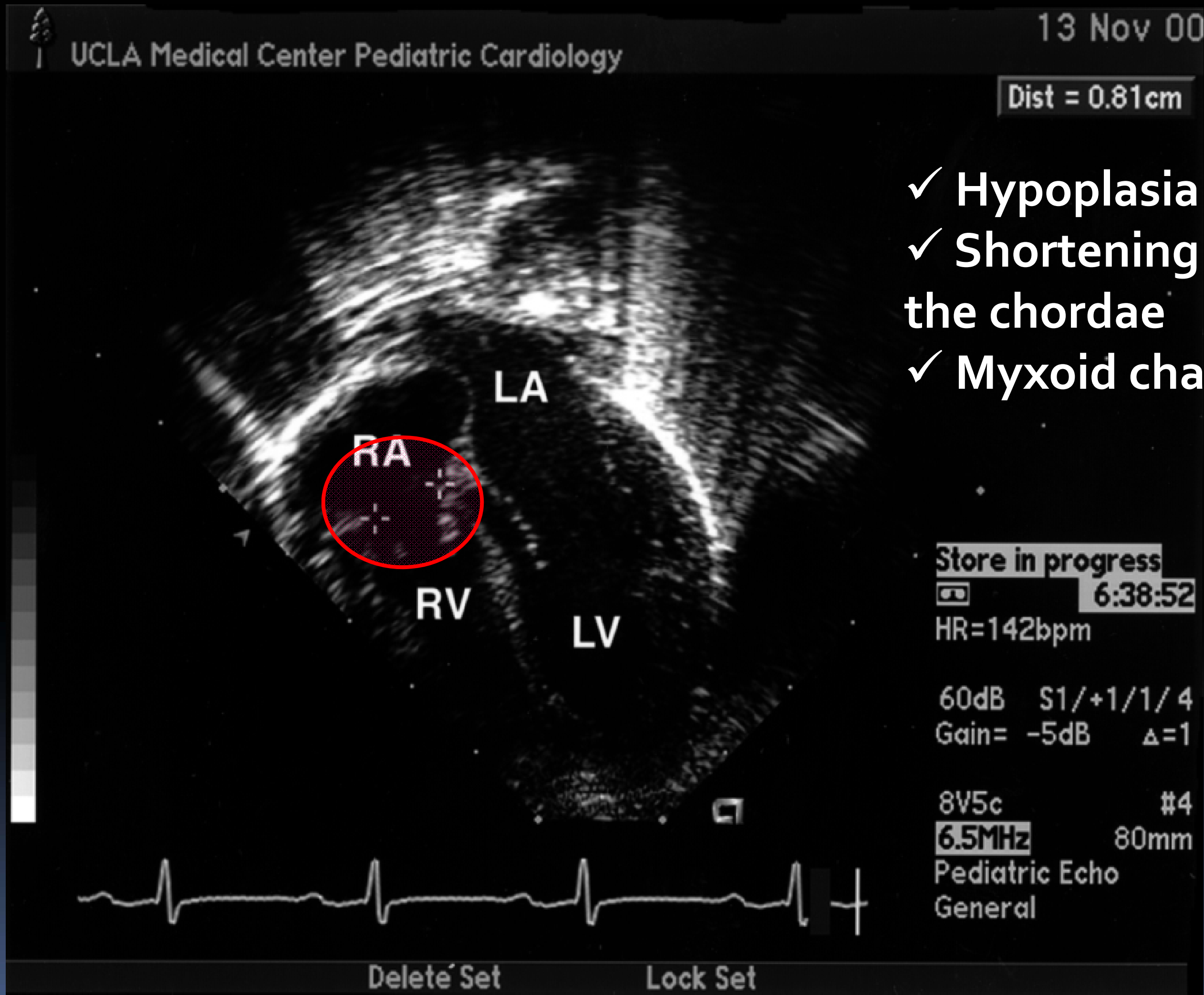
- Atretic pulmonic valve – Doppler evidence
- RV hypertrophy, small cavity
- TV size measure
- ASD (or Rt to Lt shunt PFO), PDA
- Branch PA
- Coronary circulation

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)

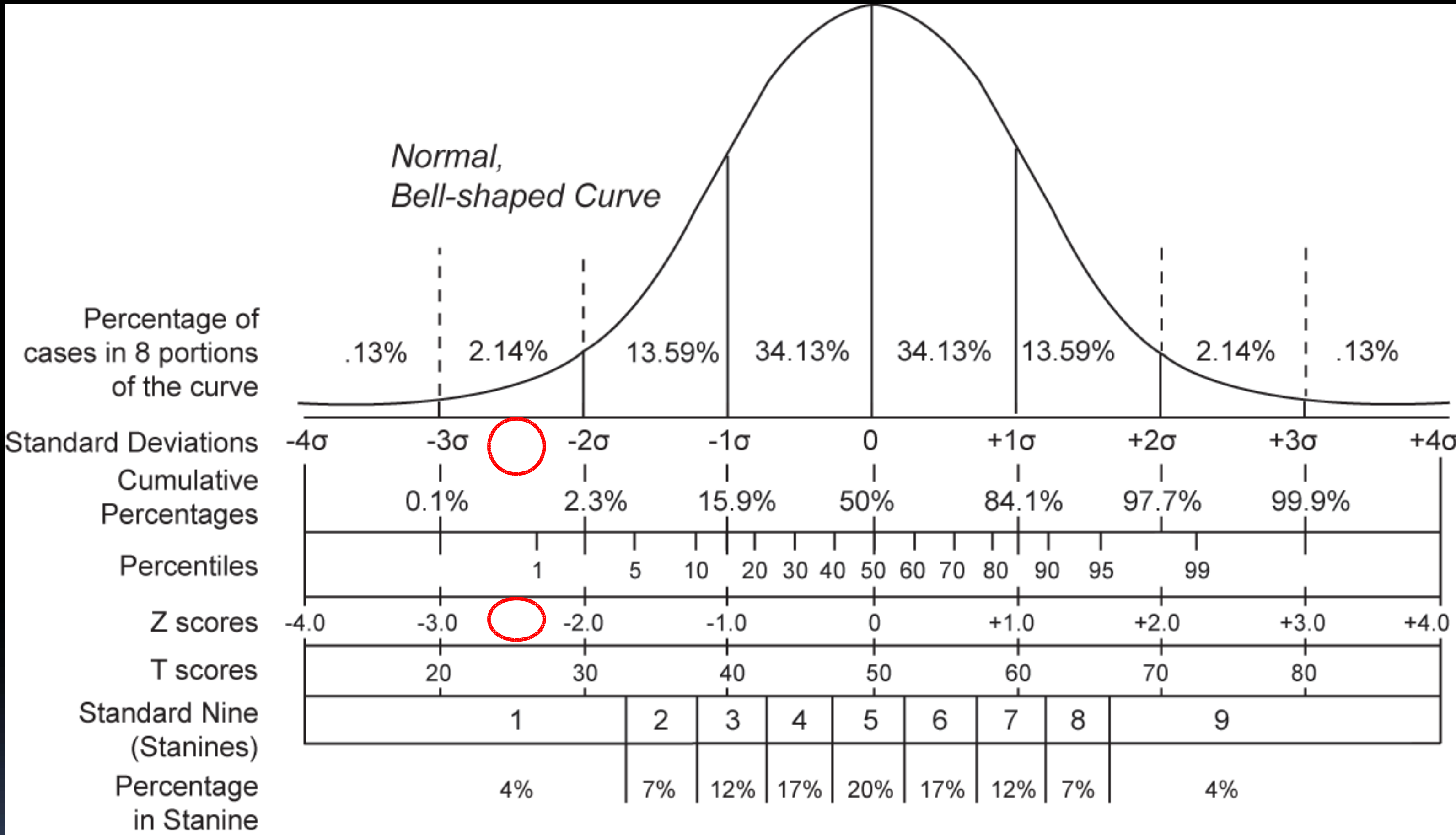
- TV annulus Z-score ; closely correlated to RV size and RV to coronary communication
- Doppler study

  ; tricuspid inflow signals – with decreasing of the color scale

  ; tricuspid insufficiency jet velocity – estimate the RV pressure

Hanley FL et al. J Thorac Cardiovasc Surg 1993
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

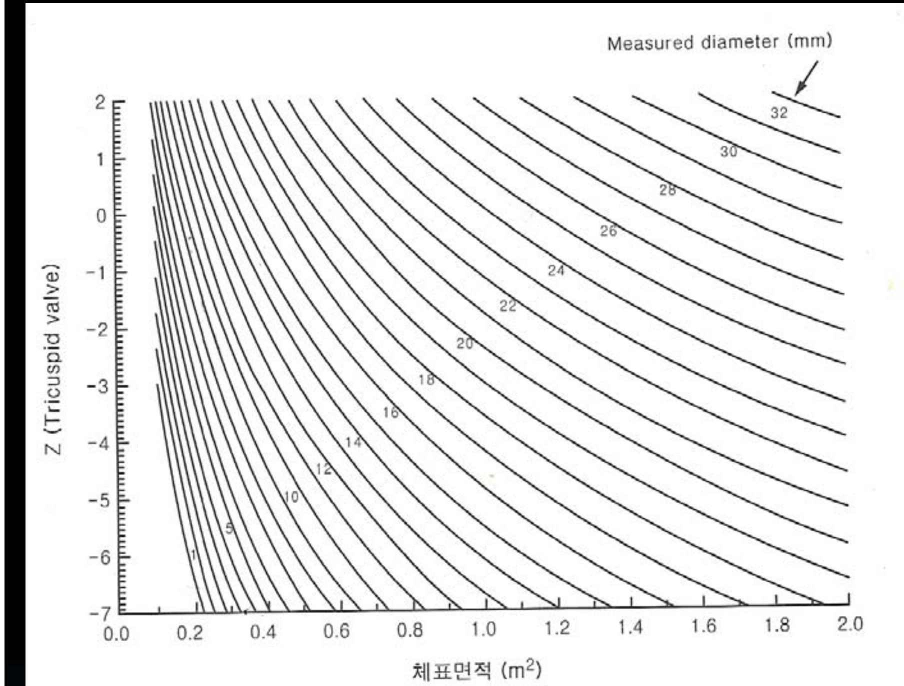
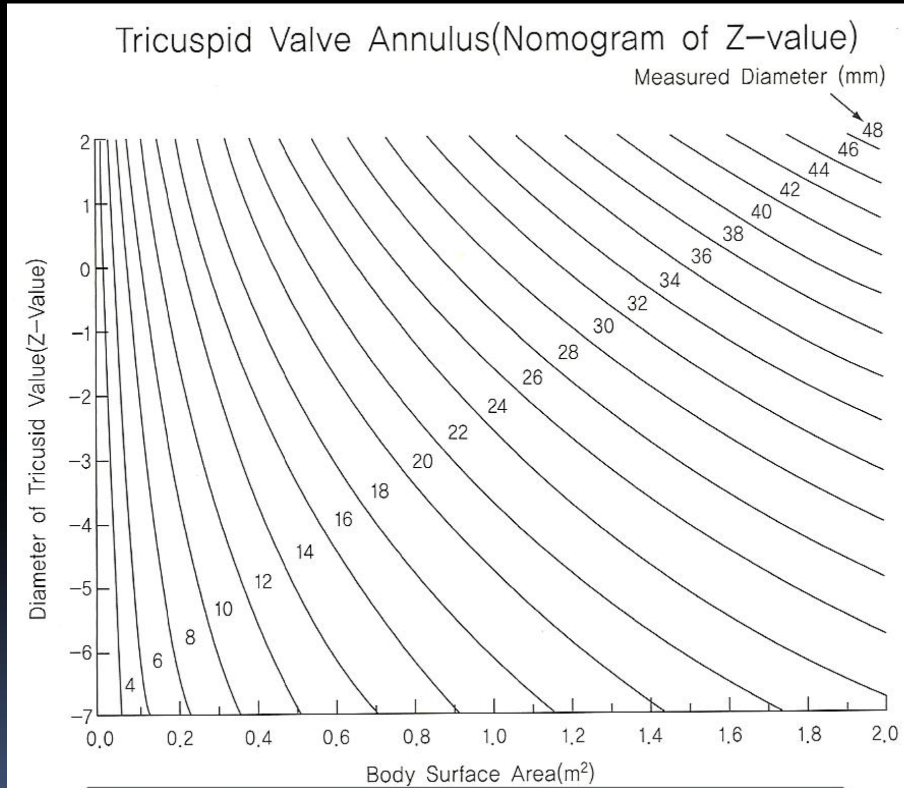


그림 1-31. 체표면적에 따른 삼첨판(tricuspid valve) 직경의 Z-value

Kirklin JW. Cardiac Surgery. 2nd ed. 1993 p30

Daubeney et al. JACC 2002



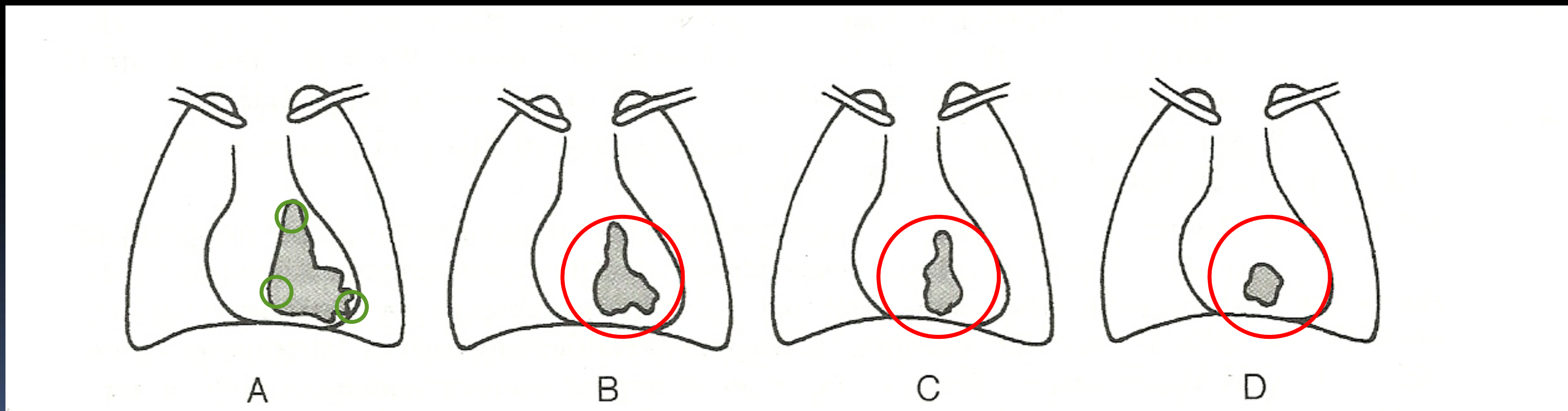
Tricuspid valve size

- TV Z-score ≥ -2.5 for BSA , tricuspid/mitral valve ratio ≥ 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

McCaffrey FM et al. J Thorac Cardiovasc Surg 1991
De Leval M et al. Circulation 1985
Bull C et al. J Thorac Cardiovasc Surg 1994

Echocardiography (RV)

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



Park MK. Pediatric Cardiology 5th ed. P 265

RV size and morphology

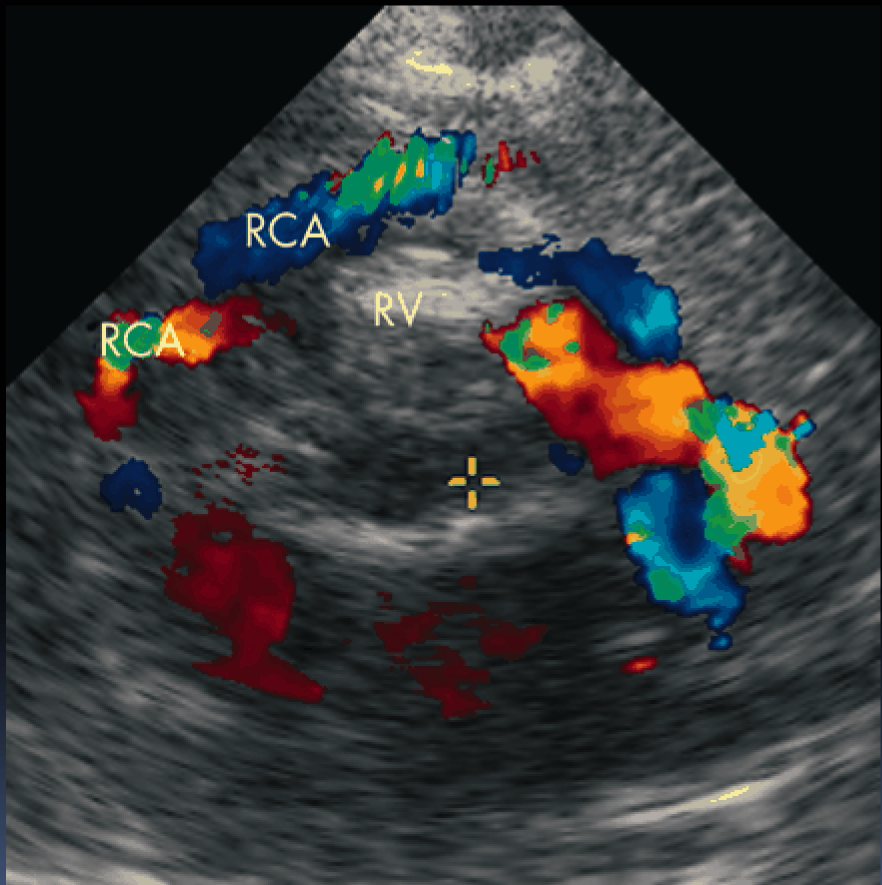
- Quantitative assessment ; RV volume in subcostal coronal and sagittal view (by biplane Simpson's rule)
- Qualitative assessment ; RV inflow portion – compare with LV (apical view)
- The patency and size of RV outflow tract ; important for RV to coronary communication and successful decompression

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)

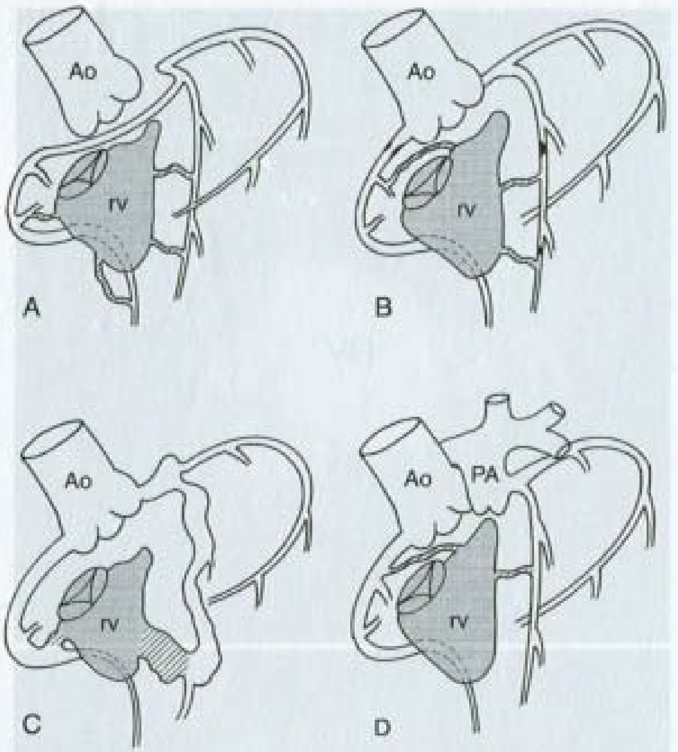
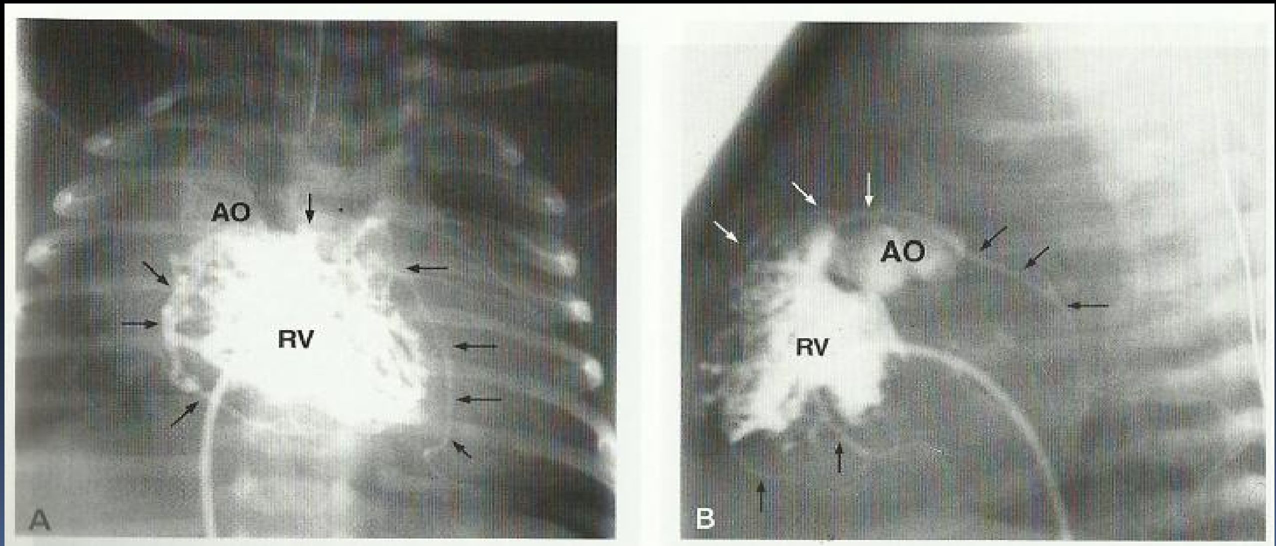


Fig. 30-4 Various forms of ventriculocoronary arterial communications causing right ventricle (RV)-dependent coronary circulation. **A**. Absent connections between the coronary arteries and aorta (Ao) with multiple ventriculocoronary connections. **B**. Proximal narrowing or interruption with multiple ventriculocoronary connections. **C**. Large fistulous communications with ectatic coronary arteries. **D**. Origin of one coronary artery from the pulmonary artery (PA) with multiple ventriculocoronary connections.

- 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

Fetal study

Echocardiographic Predictors of Outcome in Fetuses with Pulmonary Atresia with Intact Ventricular Septum

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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 *z* 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

Fetal study

Determinants of Outcome in Fetal Pulmonary Valve Stenosis or Atresia With Intact Ventricular Septum

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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 ≤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. (Am J Cardiol 2007;99:699–703)

Sick Children Hospital, University of Toronto

Catheterization

- Hemodynamics – RV, RA pressure and O_2 sat
- RV angiogram – RV morphology, size, coronary fistula
- Aortography – arch, PDA, MAPCA, coronary angio
- Balloon valvotomy – potential opening, check RV dependent coronary circulation

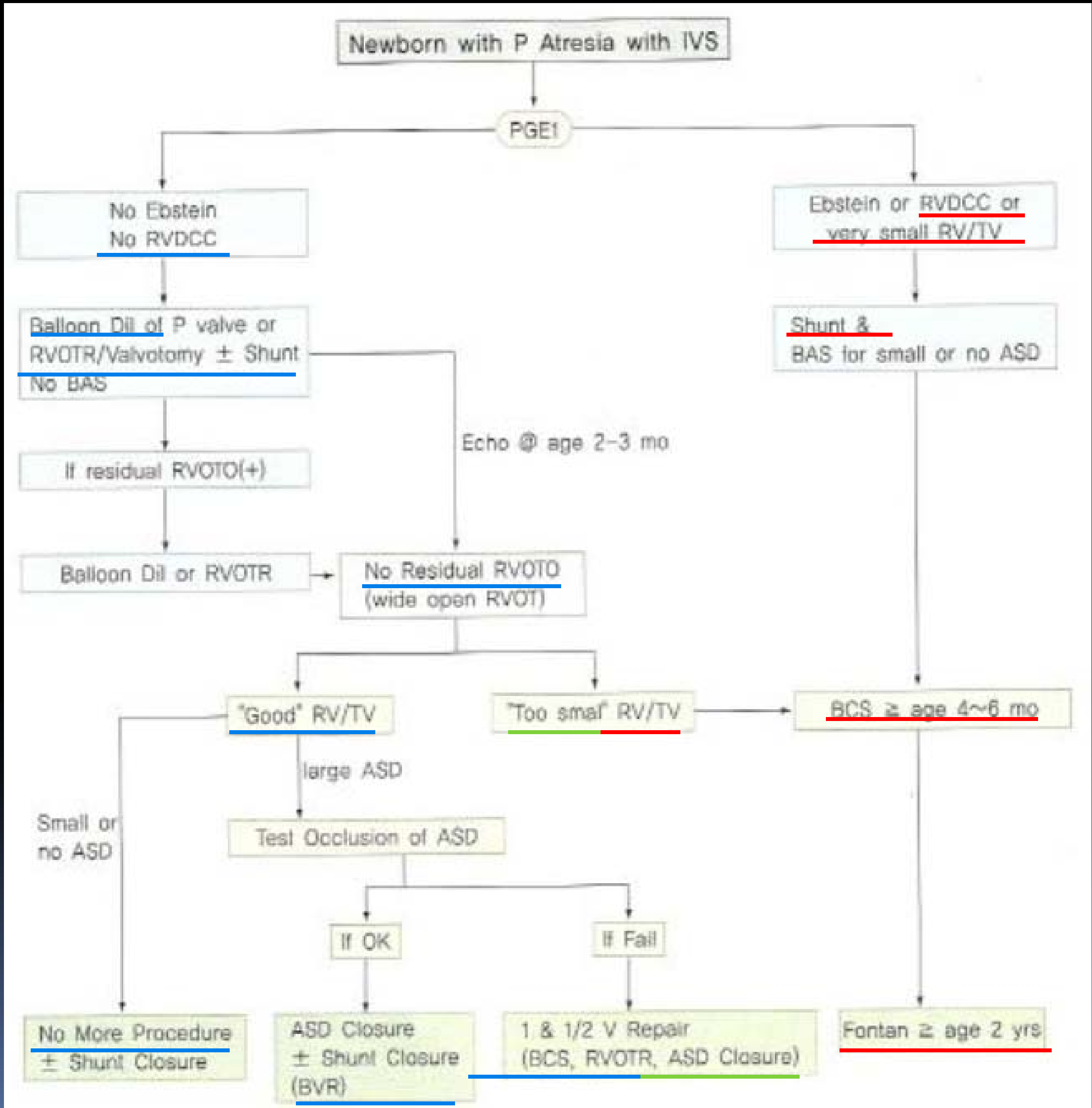
Treatment strategies

- 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

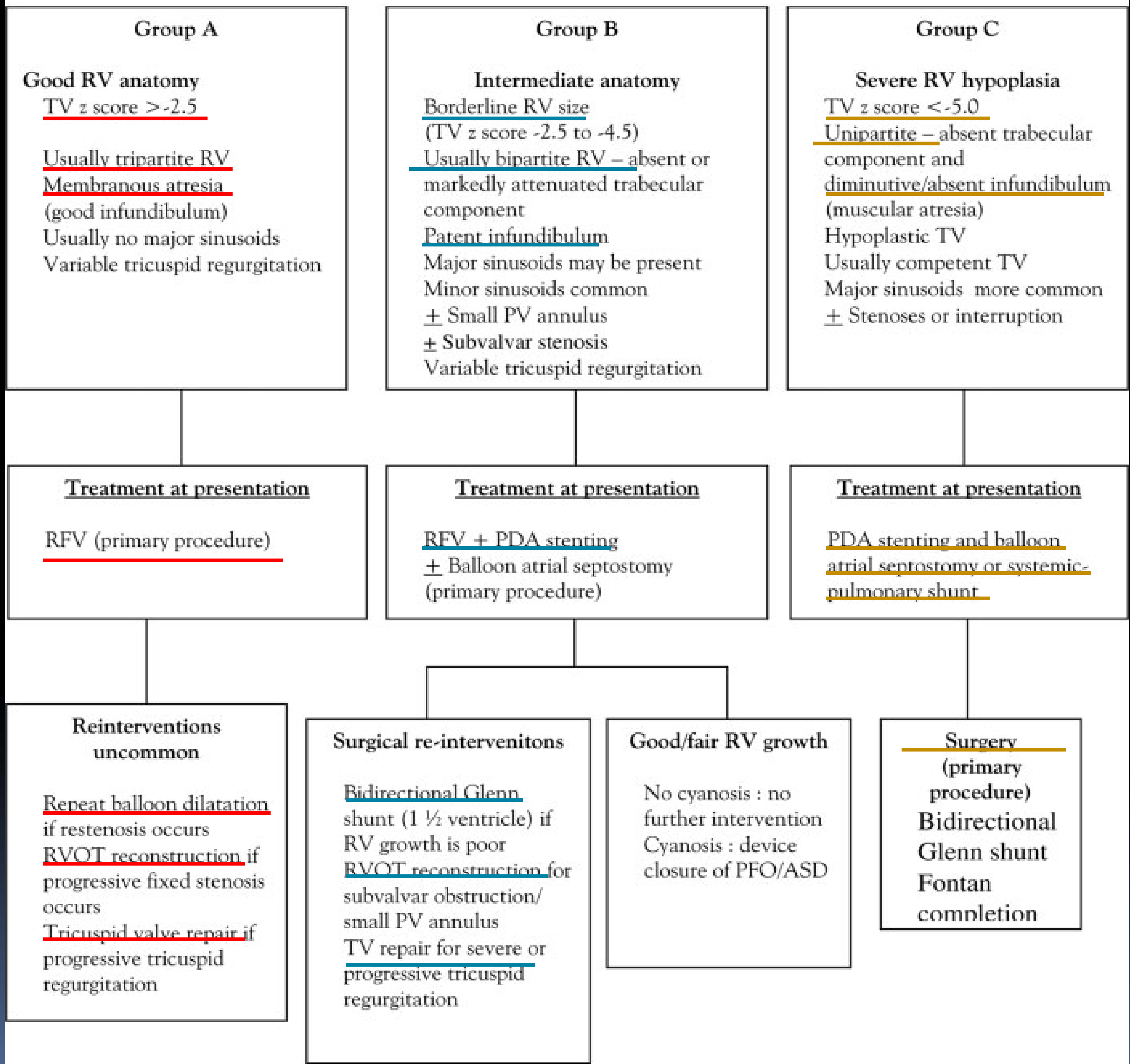
Bichell DP. Curr Opin Cardiol 1999

Treatment Algorithm



박인숙. 선천성 심장병 2nd ed. p 556

Morphologic classification and treatment strategies



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 – <u>RV end diastolic volume</u>, <u>TV annulus diameter</u>

Hanley FL et al. J Thorac Cardiovasc Surg 1993
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

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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.

(Ann Thorac Surg 2011;91:555–60)

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

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Int J Cardiol 2011

Table 1
Demographics and initial operative strategies of PA/IVS.

Characteristics (n = 20)	Value (n = 20)[a]
Age at evaluation, years[b]	28 (18–38)
Male gender	9
Initial operative strategy	
Univentricular, Fontan	7
Univentricular, palliative shunts	5
Biventricular repair	8
Initial arterial to pulmonary shunt	8
Pulmonary valvotomy, isolated	4
RV to PA conduit/RVOT reconstruction + pulmonary valvotomy	4

Table 2
Characteristics of PA-IVS cohort, surgical subgroups.

Characteristics[a]	Univentricular, palliated (n = 5)	Univentricular, Fontan (n = 7)	Biventricular repair (n = 8)	Total number (n = 20)
Average age, years[b]	29 (23–35)	28 (23–32)	30 (18–39)	29 (18–39)
Number of patients alive	4	5	6	15
NYHA class I–II[b]	4	4	6	14
Arrhythmias	3	7	6	16
Pulmonary hypertension	4	0[c]	1	5
Endocarditis	3	1	0	4
Intracardiac thrombosis	0	4	2	6
Valvular dysfunction	1	2	8	15
Protein losing enteropathy	0	2	0	2
Echocardiogram parameters				
Left ventricular EF	55% (50–65%)	51% (45–58%)	58% (50–65%)	55% (45–65%)

Int J Cardiol 2011

Types of surgical re-interventions in adulthood (≥ 18 years).

Characteristics	Number of patients	Number of procedures	NYHA class I–II pts pre/pts post[a]	Average age, years
Univentricular, palliated (n = 5)			2/4	22 (19–24)
Patients with re-interventions, total	3/5	7 (total)		
Shunt revision, surgical	1	4		
Shunt dilation, transcatheter	2	2		
Coil embolization of collaterals	1	1		
Univentricular, Fontan (n = 7)			2/5	21 (17–32)
Patients with re-interventions, total	7/7	14 (total)		
Fontan conversion	4	4		
MAZE procedure	3	3		
Mitral valve repair/replacement	2	2		
Fontan fenestration, transcatheter	1	1		
Ascending aorta replacement	1	1		
Fontan revision	1	1		
Biventricular repair (n = 8)			3/6	27 (19–38)
Patients with re-interventions, total	7/8	31 (total)		
Tricuspid valve repair/replacement	6	7		
Pulmonary valve replacement	5	7		
RV to PA conduit replacement	1	1		
RVOT reconstruction/augmentation	6	7		
Pulmonary artery intervention, transcatheter	3	3		
Mitral valve repair/replacement	2	4		
MAZE procedure	2	2		

Long term follow up evaluation for PA c IVS
– echocardiography, EKG, Holter, MRI