

Long Term postop Follow up for the Complex Two Ventricle Repair (Rastelli operation in TGA patients)

So-Ick Jang

Department of Pediatric cardiology, Sejong Heart
Institute

Sejong General Hospital

Rastelli operation for TGA VSD with LVOTO

- Introduced in 1969
- Alternative
- 1982 REV (réparation a l'étage ventriculaire) Lecompte
- 1984 Nikaidoh (aortic root translocation)

Long-term results of Rastelli operation

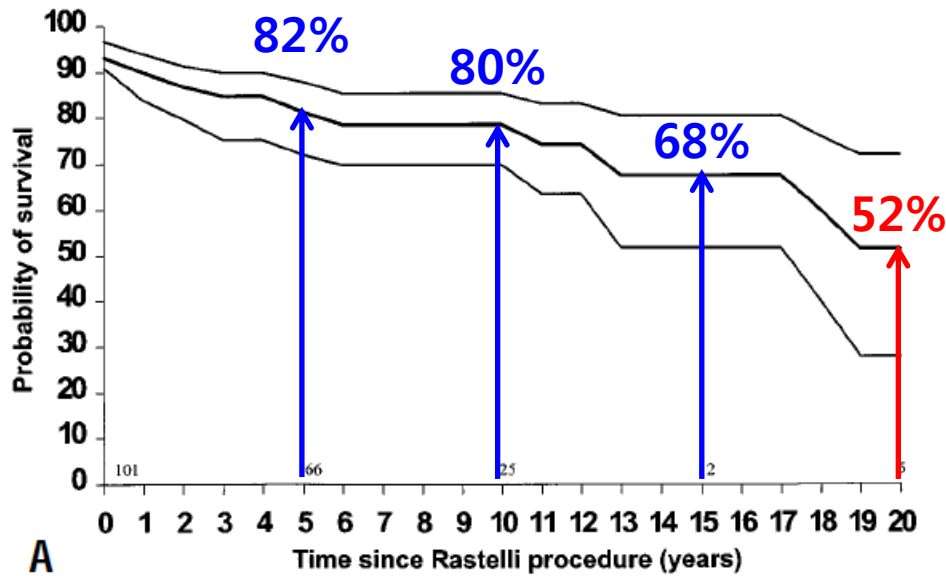


Table IV. Causes of late death or failure

Causes	n
Sudden death	5
Left ventricular dysfunction	7
Conduit pseudointima rupture	1
Myocarditis	1
Unknown	2
At reoperation	2

Kreutzer et al. TWENTY-FIVE-YEAR EXPERIENCE WITH RASTELLI REPAIR FOR TRANSPOSITION OF THE GREAT ARTERIES. *The Journal of Thoracic and Cardiovascular Surgery*, 2000

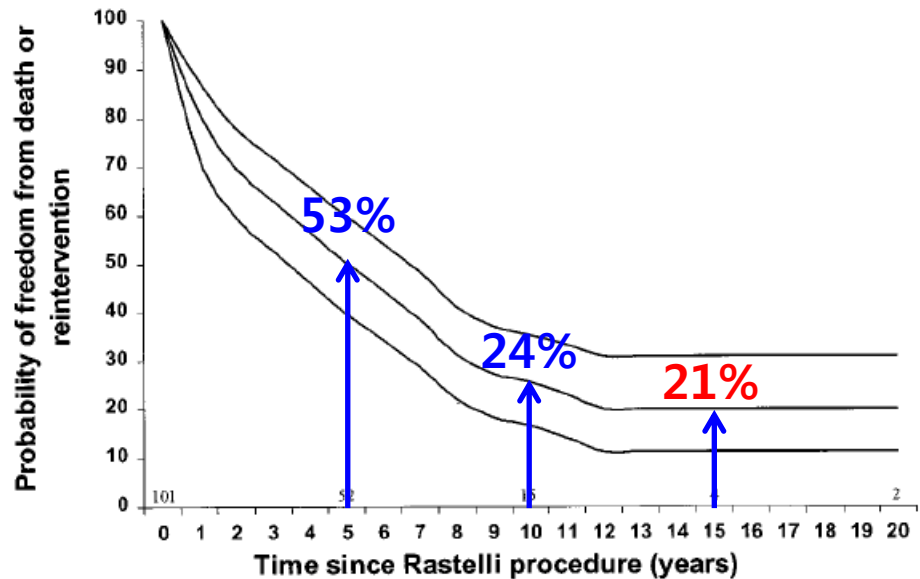
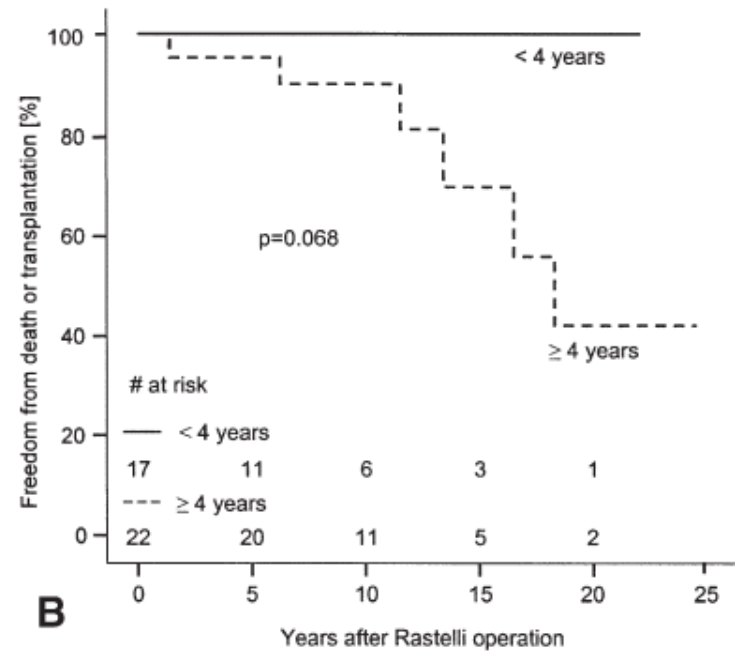
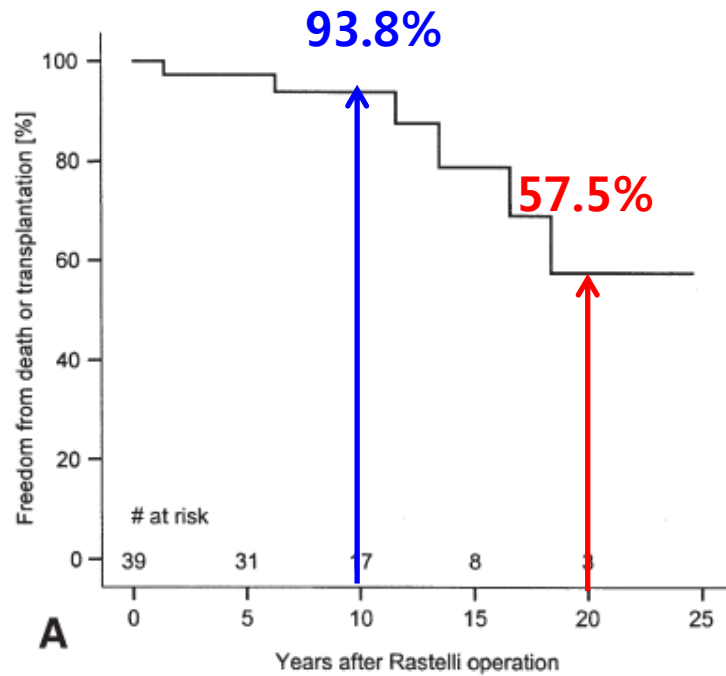


Fig 2. Freedom from reintervention, reoperation, or death.



HÖRER et al. Long-Term Results After the Rastelli Repair for Transposition of the Great Arteries. Ann Thorac Surg. 2007

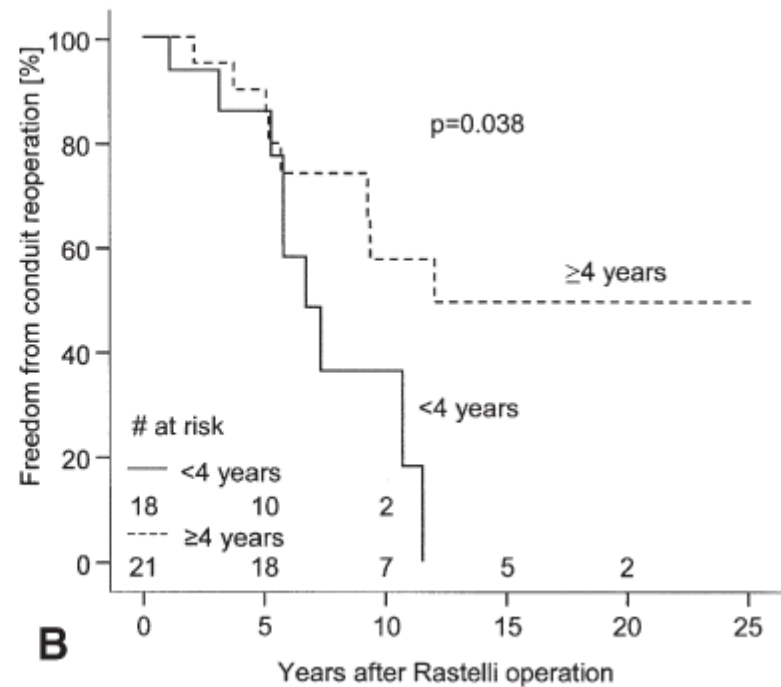
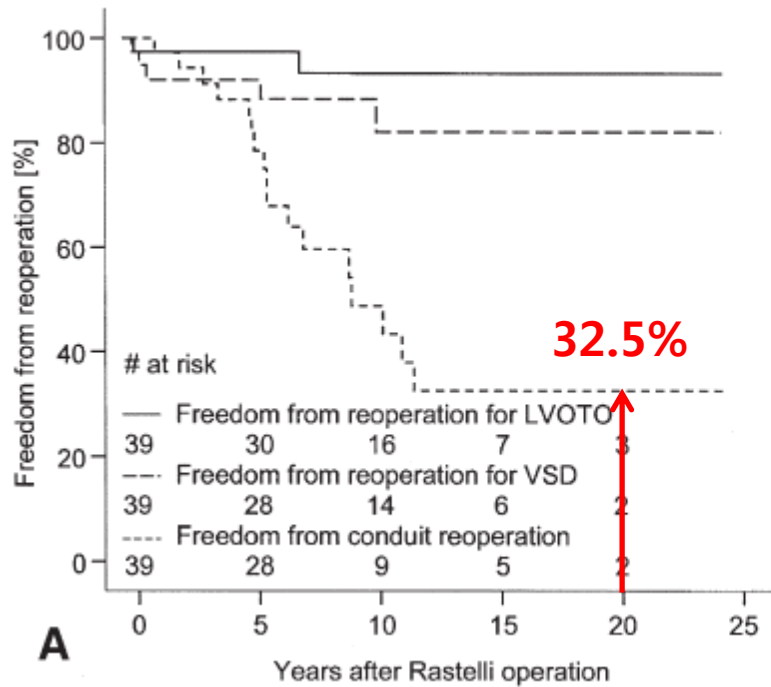


Table 4. Predictors for Death and Transplantation

Variable	Numbers	Events	<i>p</i> Value
LVOTO subvalvular and valvular	26	6	0.012
Stenosis of main pulmonary arteries within hilar bifurcation	8	2	0.018
Stenosis of peripheral pulmonary arteries	5	2	<0.001
Hemoglobin level >18.5 g/dL	19	6	0.094
Age at time of Rastelli operation >4 years	22	6	0.068
Body weight at time of Rastelli operation >17 kg	23	6	0.071
VSD enlargement	23	5	0.030
Cardiopulmonary bypass time >136 minutes	19	5	0.029
Aortic cross-clamp time >83 minutes	19	6	0.015

LVOTO = left ventricular outflow tract obstruction; VSD = ventricular septal defect.

Rastelli Operation for Transposition of the Great Arteries With Ventricular Septal Defect and Pulmonary Stenosis

John W. Brown, MD, Mark Ruzmetov, MD, PhD, Daniel Huynh, BS,
Mark D. Rodefeld, MD, Mark W. Turrentine, MD, and Andrew C. Fiore, MD

Section of Cardiothoracic Surgery, Indiana University School of Medicine, Indianapolis, Indiana; and St. Louis University School of Medicine, St. Louis, Missouri

Rastelli Operation for Transposition of the Great Arteries With Ventricular Septal Defect and Pulmonary Stenosis

John W. Brown, MD, Mark Dargatzis, MD, PhD, Daniel H. Frank, BS

Background. The optimal surgical treatment of patients with transposition of the great arteries, ventricular septal defect, and pulmonary stenosis is controversial. Although the Rastelli operation has been standard surgical management of this lesion, aortic root translocation with right ventricular outflow tract (RVOT) reconstruction (Nikaidoh) and the pulmonary artery translocation (Lecompte) or REV (réparation à l'étage ventriculaire) are surgical alternatives more recently introduced to treat this complex lesion. This report reviews our 20-year experience with the Rastelli procedure and attempts to compare our outcomes with those recently published using the Nikaidoh and REV procedures.

Methods. Between 1988 and 2008, 40 patients (median age, 4 years; range, 9 months to 17 years) underwent Rastelli operation at our institutions. The RVOT was obstructed in 32 and atretic in 8. Follow-up was available for all but one patient (mean follow-up, 8.6 ± 5.6 years). The RVOT was reconstructed with homograft ($n = 25$), bovine jugular vein ($n = 8$), nonvalved Dacron tube ($n = 5$), or a porcine valved conduit ($n = 2$). Two patients required a pacemaker.

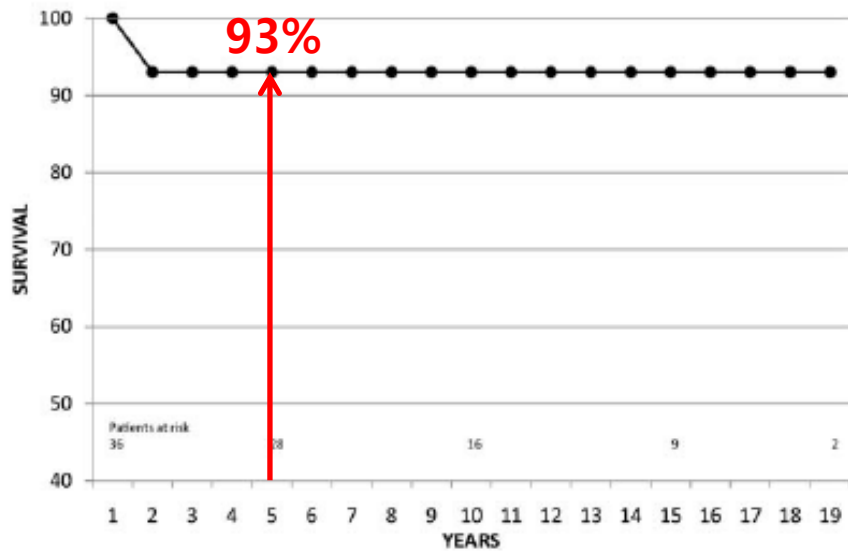
Results. There were no early, but three late deaths and one heart transplantation 12 years postoperative the Rastelli operation. Kaplan-Meier survival was 93% at 5, 10, and 20 years. Univariate risk factors for death or trans-

plantation included surgery before 1998 ($p = 0.03$) and concomitant noncardiac anomalies ($p = 0.001$). Sixteen patients (40%) had reoperation for right ventricular-pulmonary artery conduit stenosis (mean, 7.8 ± 3.8 years) without mortality. Freedom from conduit replacement was 86%, 74%, 63%, and 59% at 5, 10, 15, and 20 years, respectively. Multivariate analysis revealed that the risk factors of conduit replacement were younger age at operation ($p = 0.001$) and surgery before 1998 ($p < 0.001$). Two patients (5%) required reoperation for left ventricular outflow tract obstruction. At follow-up, there were no sudden unexplained deaths, and New York Heart Association functional class is I or II.

Conclusions. The Rastelli procedure is a low-risk operation with regard to early and late mortality and reoperation for left ventricular outflow tract obstruction. Conduit change operations will be required in most patients regardless of the technique of repair, but currently can be performed with low morbidity and mortality. These midterm outcomes after the Rastelli operation should serve as a basis for comparison with surgical alternatives more recently introduced for transposition of the great arteries and ventricular septal defect with RVOT obstruction.

(Ann Thorac Surg 2011;91:188–94)

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Kaplan-Meier survival was 93% at 5, 10, and 20 years. Freedom from death or cardiac transplantation was 92%±4% and 90%±5% at 10 and 20 years, respectively.

Fig 1. Survival curve in patients with Rastelli operation.

Freedom from reoperation for LVOTO or conduit change at 20 years was 95%±2 % and 60 %±6.4%, respectively (Fig 2).

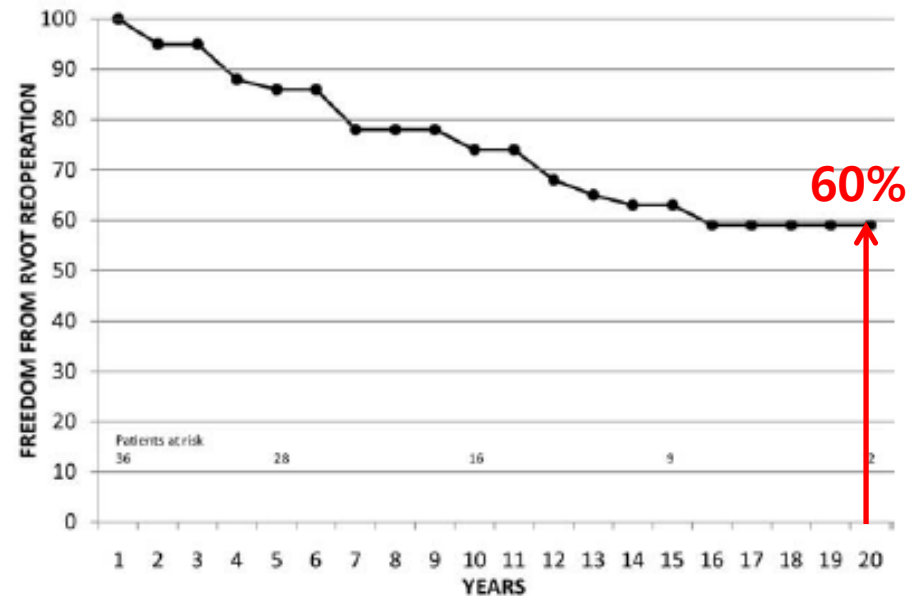


Fig 2. Freedom from reoperation curve in patients with Rastelli operation.

Cause of death

- Sudden Cardiac death
- Heart Failure
- Reoperation

Late arrhythmia

- Cause by
- RBBB → bifascicular & complete AV block
- scar tissue
- RV hypertension due to an obstructed conduit

- AV block
- Supraventricular tachycardia
- Ventricular tachycardia

Left ventricular failure

- Remained LV dilatation & hypertrophy
- Prolonged hypoxia, obstruction, volume overloading for age of operation
- Abnormal ventricular septum with a large prosthetic components(VSD baffle)
- Right ventricular hypertension→ impairing LV filling & leftward septal displacement

Follow Up

Echocardiography

- the 1st-line diagnostic technique
- LV and RV function : Functional study using 3D
Doppler tissue imaging
Tei index, E/E'
- LV→AV
- Function of the conduit between the RV and PA
RVP from TR velocity, IVS shape
- Residual VSDs

Cardiac MR

- 2nd method when Echo measurements are borderline or ambiguous : function, volume, valve regurgitation
- 3D anatomical reconstruction
- not restricted by body size or acoustic windows
- A detection and quantification of myocardial fibrosis/scar (gadolinium late enhancement)
- for volumetric measurements, assessment of vessels, and detection of myocardial fibrosis.

Computed Tomography (CT)

- excellent spatial resolution and rapid acquisition time.
- ECG-gated image
- Particularly good for imaging epicardial C.A. and collateral arteries, and for parenchymal lung disease.
- Ventricular size and function (inferior to CMR)
- require staff with expertise in complex CHD.

Cardiopulmonary exercise test

- Quality of life and functional capacity
- Objective exercise capacity (time, VO_{2max}),
Ventilation efficiency (V_E/V_{CO2} slope),
Chronotropic and BP response,
Exercise-induced arrhythmia
- Serial exercise testing

Cardiac catheterization

- **For Diagnosis (Class IIa)**
- Coronary artery delineation before any intervention for RVOTO (Level of Evidence: C)
- Assessment of residual VSD. (Level of Evidence: C)
- Assessment of PAH, with potential for vasodilator testing. (Level of Evidence: C)
- Assessment of subaortic obstruction across the LV-to-Ao. tunnel. (Level of Evidence: C)

Cardiac catheterization

- LV and RV diastolic function, Pressure gradient, shunt quantification and the evaluation of extracardiac vessels such as aortic pulmonary collateral arteries.
- Measurement of oxygen uptake rather than estimation
- CAG in men ≥ 40 years of age, postmenopausal women, and patients with signs of or risk factors for CAD.

Cardiac catheterization

- **For Intervention (Class IIa)**
- Dilation with or without stent implantation of conduit obstruction ($p(RV/Ao) \geq 50\%$, or peak-to-peak gradient greater than 30 mm Hg; these indications may be lessened in the setting of RV dysfunction).
(Level of Evidence: C)
- Device closure of residual VSD. (Level of Evidence: C)

Surgical Intervention

- **Class I**

Conduit obstruction peak-to-peak gradient ≥ 50 mm Hg.

(Level of Evidence: C)

RV/LV pressure ratio ≥ 0.7 . (Level of Evidence: C)

Lesser degrees of conduit obstruction if pregnancy is being planned or greater degrees of exercise are desired.

(Level of Evidence: C)

Subaortic (baffle) obstruction (mean gradient ≥ 50 mm Hg).

(Level of Evidence: C)

Lesser degrees of subaortic (baffle) obstruction if LV hypertrophy is present, pregnancy is being planned, or greater degrees of exercise are desired. (Level of Evidence: C)

Presence of concomitant severe AR. (Level of Evidence: C)

Surgical Intervention

- **Class I**

for conduit regurgitation (severe)

Symptoms or declining exercise tolerance.

(Level of Evidence: C)

Severely depressed RV function. (Level of Evidence: C)

Severe RV enlargement. (Level of Evidence: C)

Development/progression of atrial or ventricular arrhythmias. (Level of Evidence: C)

More than moderate TR. (Level of Evidence: C)

Surgical Intervention

- **Class I**

For Surgical closure of residual VSD

Qp/Qs greater than 1.5:1. (Level of Evidence: B)

Systolic PA pressure \geq 50 mm Hg. (Level of Evidence: B)

Increasing LV size from volume overload. (Level of Evidence: C)

Decreasing RV function from pressure overload.

(Level of Evidence: C)

RVOT obstruction (peak instantaneous gradient \geq 50 mm Hg).

(Level of Evidence: B)

PA pressure \leq 2/3 of systemic pressure, or PVR \leq 2/3 of SVR, with a net left-to-right shunt of 1.5:1, or a decrease in PA pressure with pulmonary vasodilators (oxygen, nitric oxide, or prostaglandins). (Level of Evidence: B)

Arrhythmias

- Risk stratification, investigation, and choice of Tx. are often different from those applied to the normally formed heart.
- onset of arrhythmias
→ signal of hemodynamic decompensation
- Catheter ablation should be considered when symptomatic tachyarrhythmias require action and interventional treatment is feasible.
- Antiarrhythmic drug therapy?

Electrophysiology Testing/Pacing

- **Class I**

Pacemaker implantation is recommended for patients with d-TGA with either symptomatic sinus bradycardia or sick sinus syndrome. (Level of Evidence: B)

- **Class IIa**

Routine surveillance with history, ECG, assessment of RV function, and periodic Holter monitoring can be beneficial as part of routine follow-up.

(Level of Evidence: B)

Sudden cardiac death

- Unexplained syncope is an alarming event.
- ICD implantation is indicated in survivors of cardiac arrest after exclusion of reversible causes (IB).
- In the absence of a defined and reversible cause, ICD implantation is reasonable (IIaB).
- Recommended therapy includes catheter ablation or surgical resection to eliminate VT. If that is not successful, ICD implantation is recommended.

Sudden cardiac death

- Invasive hemodynamic and EP evaluation is reasonable in patients with unexplained syncope and impaired ventricular function.
- Patients with spontaneous sustained VT should undergo invasive hemodynamic and EP evaluation.
- EP testing may be considered for patients with ventricular couplets or non-sustained VT to determine the risk of sustained VT (IIbC).
- Prophylactic antiarrhythmic therapy is not indicated for asymptomatic patients (IIIc).

Pregnancy

- Patients with hemodynamic compromise have elevated risks during pregnancy; increased risk of developing **life-threatening arrhythmias, or RV dysfunction.**
- Patients who are in good clinical condition have a high likelihood of going through a **normal pregnancy**, although the risks of miscarriage, fetal growth retardation, and early delivery are higher than in the normal population.

Case

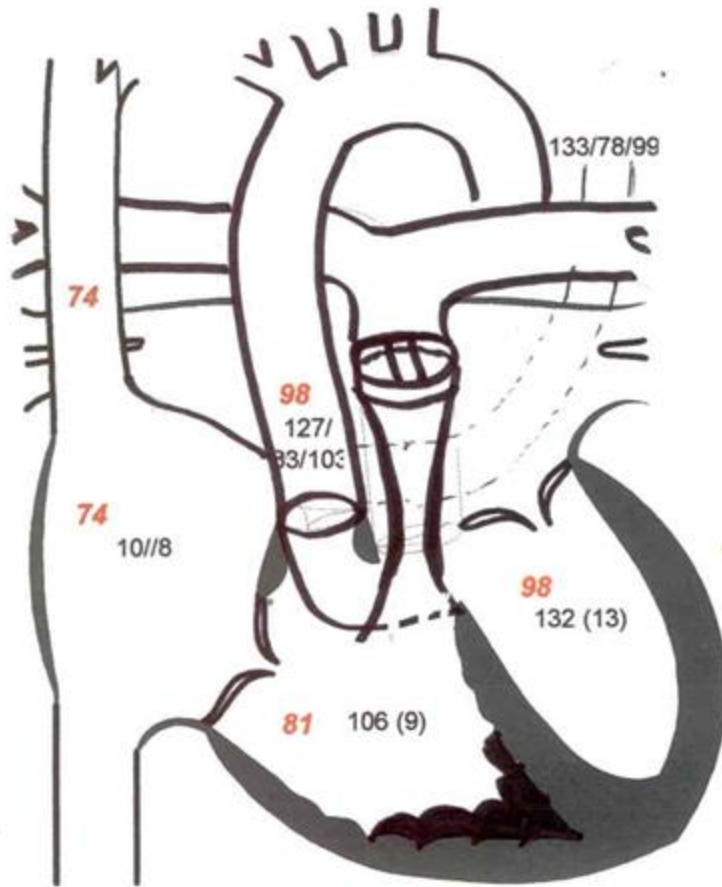
- 30yr male
- Dx> TGA, VSD, PS
- PHx>

Rastelli operation(valved conduit 23mm), VSD widening
(1990-05-30)

RVOT widening with patch of Gore-Tex graft 24mm
(2003-3-12)



2003-01-13, 19years old



Diagnosis/Intervention

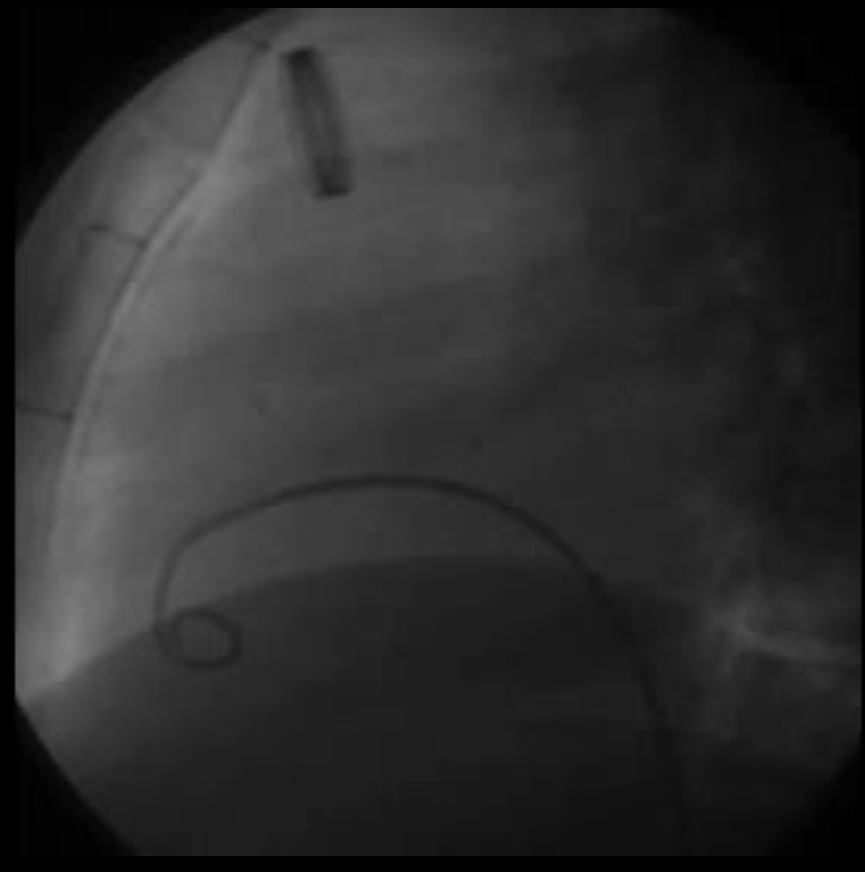
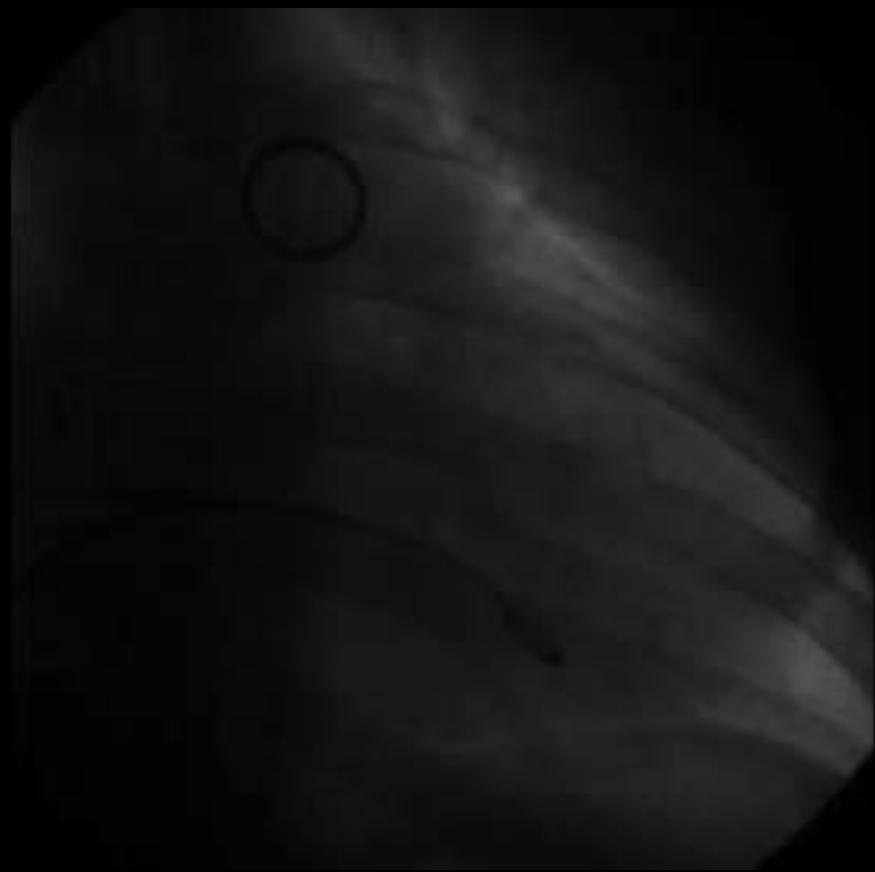
s/p Total correction (Rastelli op with CM23mm)(90-5-30)

DORV or TGA with multiple large VSDs, PS, bilat SVC, LSVC to CS

<Findings>

1. Severe conduit stenosis
acceptable PV function
2. Severe RVH
acceptable RV function
3. No LVOTO
4. Original MPA flow ~~low~~ → dead space
5. AR G I
TR trivial

p(RV/Ao) : 0.83



ID: **1985-00711**
Age: **30 yr**
Gender: **Male**
Date of Birth:

Hookup Date: **21-Jan-2014**
Hookup Time: **09:25:00**
Duration: **23:10:00**
Recorder Serial #: **8208d107**
Order #:

Overreading Physician: ,
Referring Physician: ,
Ordering Physician: ,
Hook-Up Technician: **Park , M.H**
Analyzing Technician: ,
Indication/Diagnosis: **Dr. Jang so ik**
Medications: **Jang PED**

General

91494 QRS complexes
74 Ventricular beats (< 1%)
31 Supraventricular beats (< 1%)
< **1** % of total time classified as noise

Ventriculars (V, F, E, I)

72 Isolated
1 Couplets
0 Bigeminal cycles
0 Runs totaling 0 beats

Heart Rates

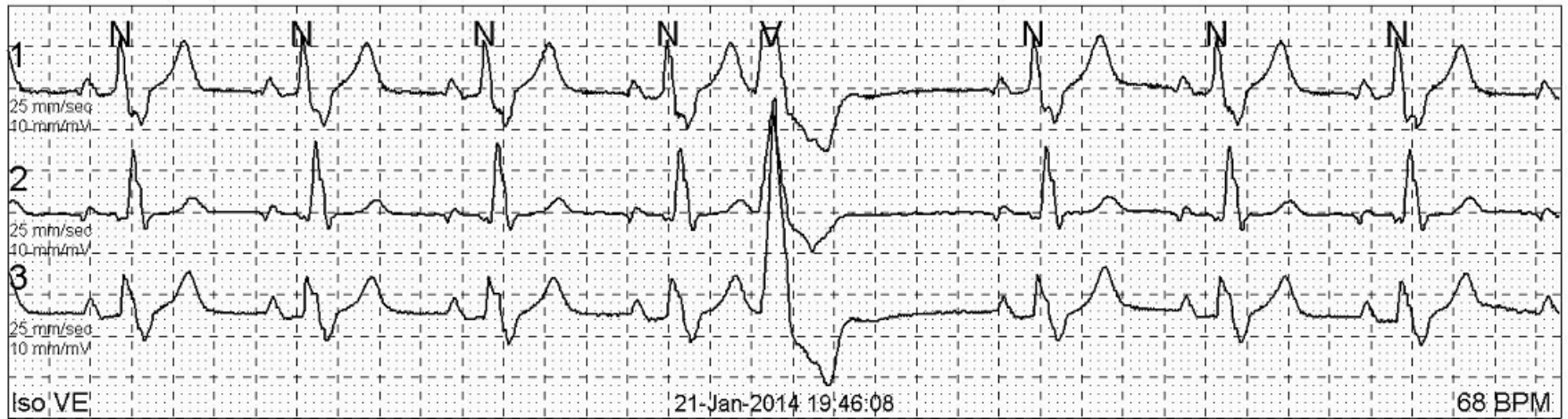
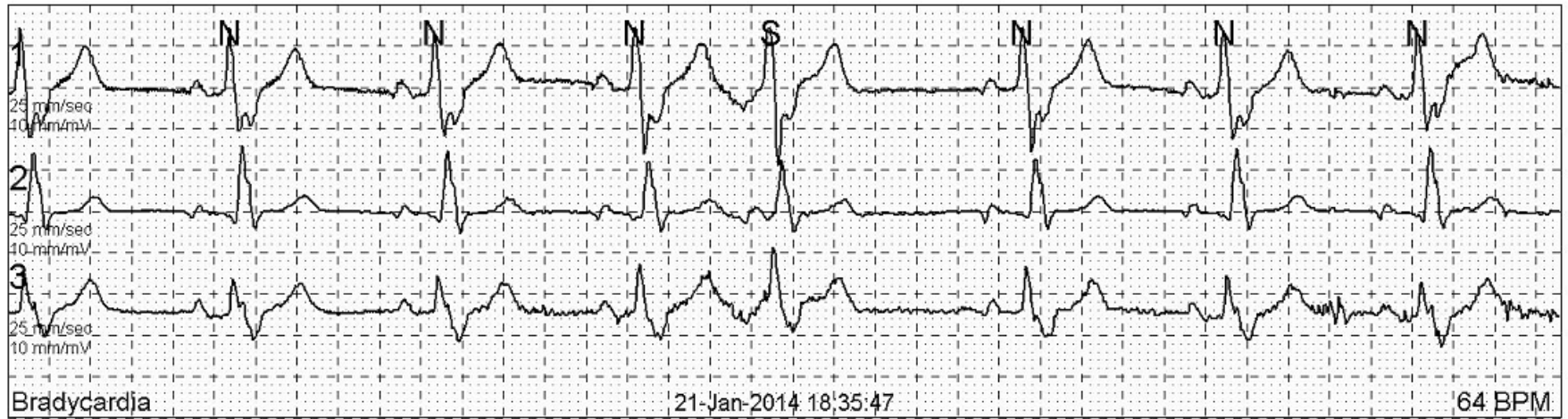
54 Minimum at 12:21:21 21-Jan
66 Average
94 Maximum at 19:52:14 21-Jan
0 Beats in tachycardia (≥ 100 bpm), 0% total
10071 Beats in bradycardia (≤ 60 bpm), 11% total
1.5 Seconds Max R-R at 07:54:01 22-Jan

Supraventriculars (S, J, A)

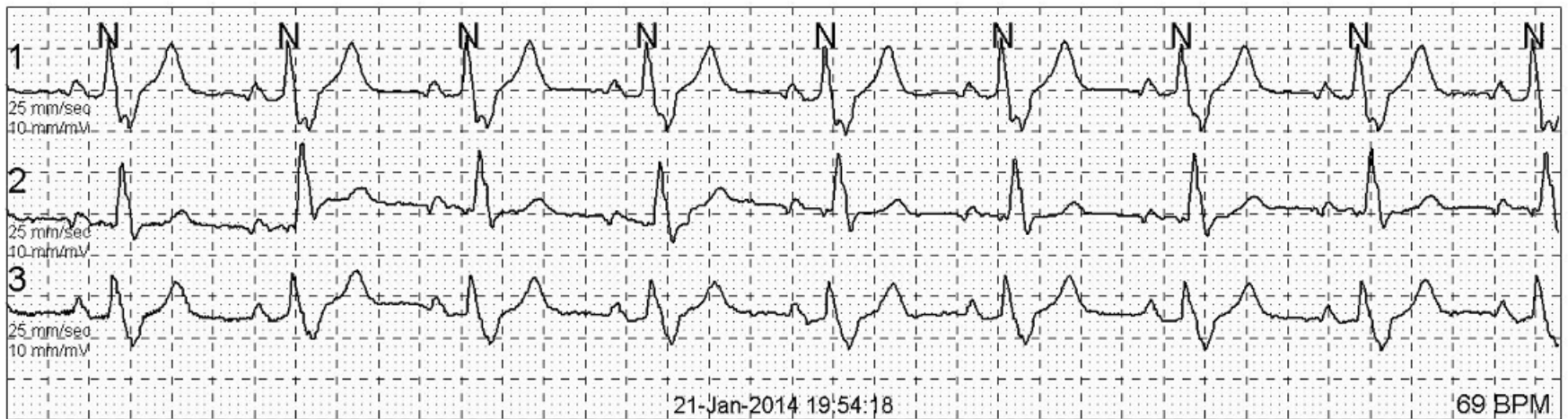
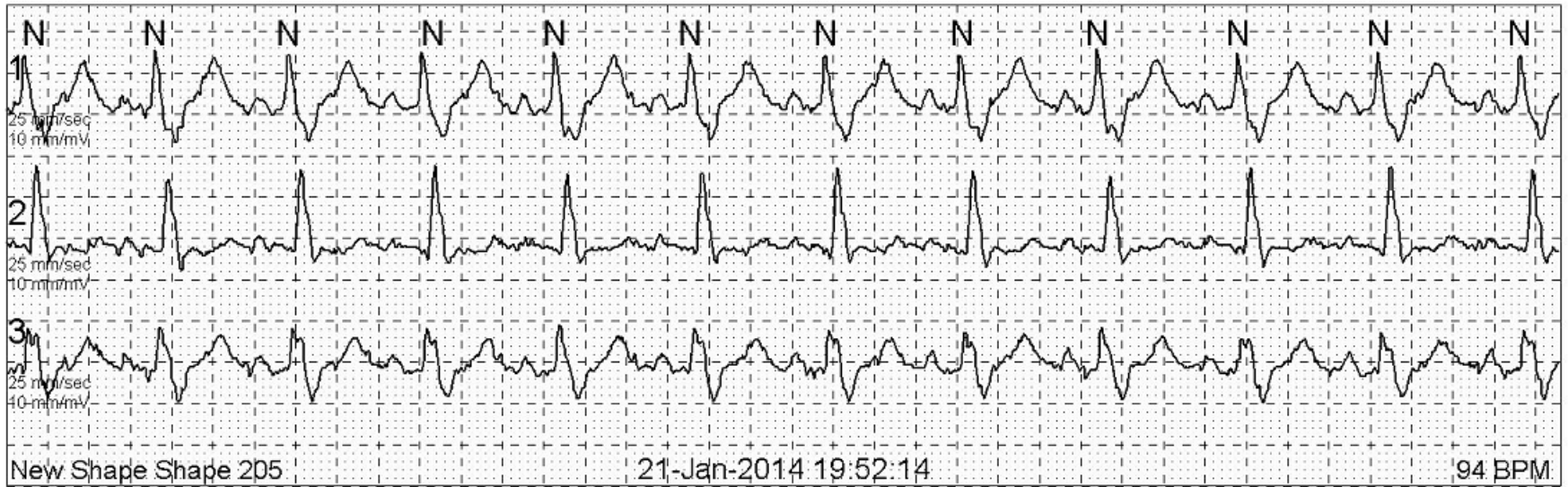
29 Isolated
1 Couplets
0 Bigeminal cycles
0 Runs totaling 0 beats

ID: 1985-00711

Location: Unknown
Hookup: 21-Jan-2014

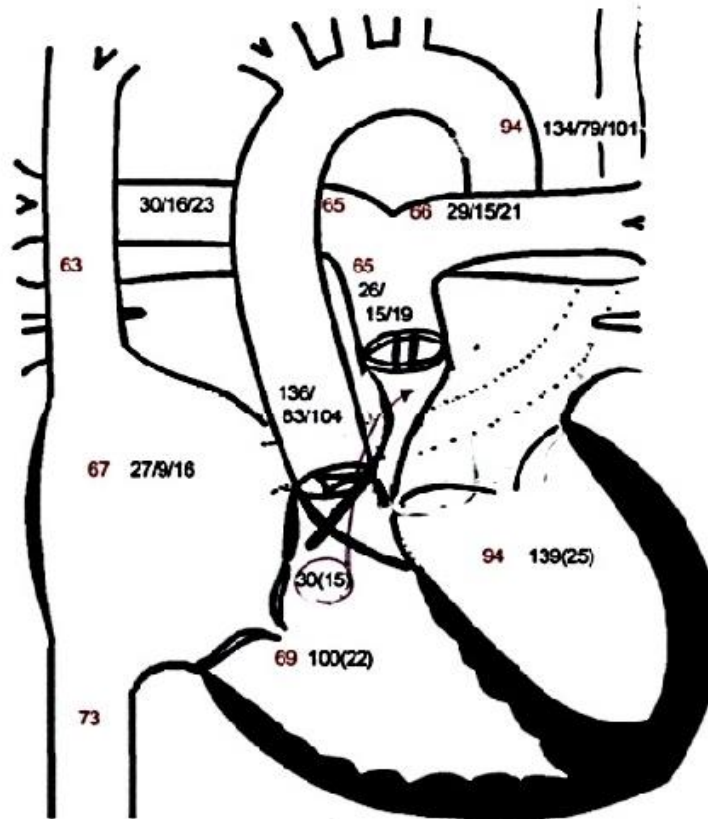


Max Heart Rate



ABGA : 7.406-44.6-81.3-

(Sat : 96.2%)



Diagnosis/Intervention

s/p Rastelli op (1990-05-30, 6y10m)
s/p RVOT widening with patch of Gore-Tax
graft 24mm (200-03-12, 19yr)
s/p Hematoma removal (2003-03-13)

{S,D,D}, Left arch
DORV(TGA type), VSD, PS, bilateral SVC

Findings>

1. severe conduit stenosis, PG=70mmHg
narrowest d=8mm
2. valve function : OK
3. high both VEDP
LVEDP=25mmHg, RVEDP=22mmHg
4. Ventricular function : Fair

Qp/Qs: 1

Qp : 1.9

Qs : 1.9

TPG : 1

Rp : 0.5

Rs : 46.3

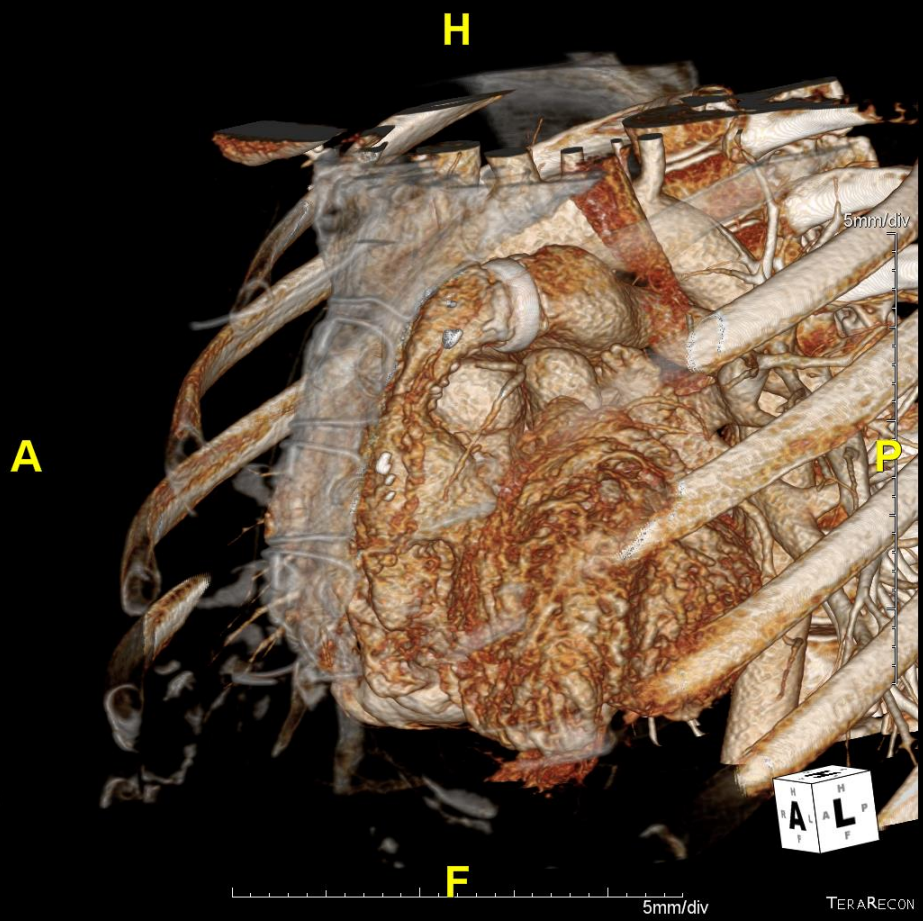
p(RV/Ao): 0.66

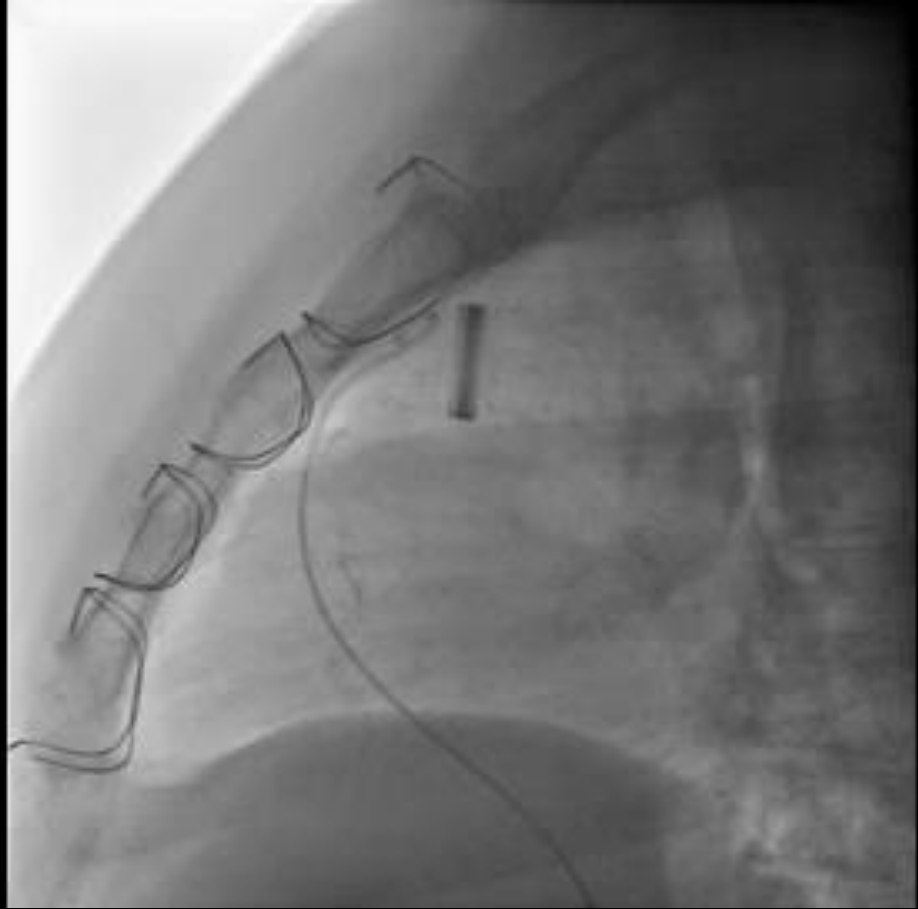
RVO/Ao=30/15 (152/88/113)

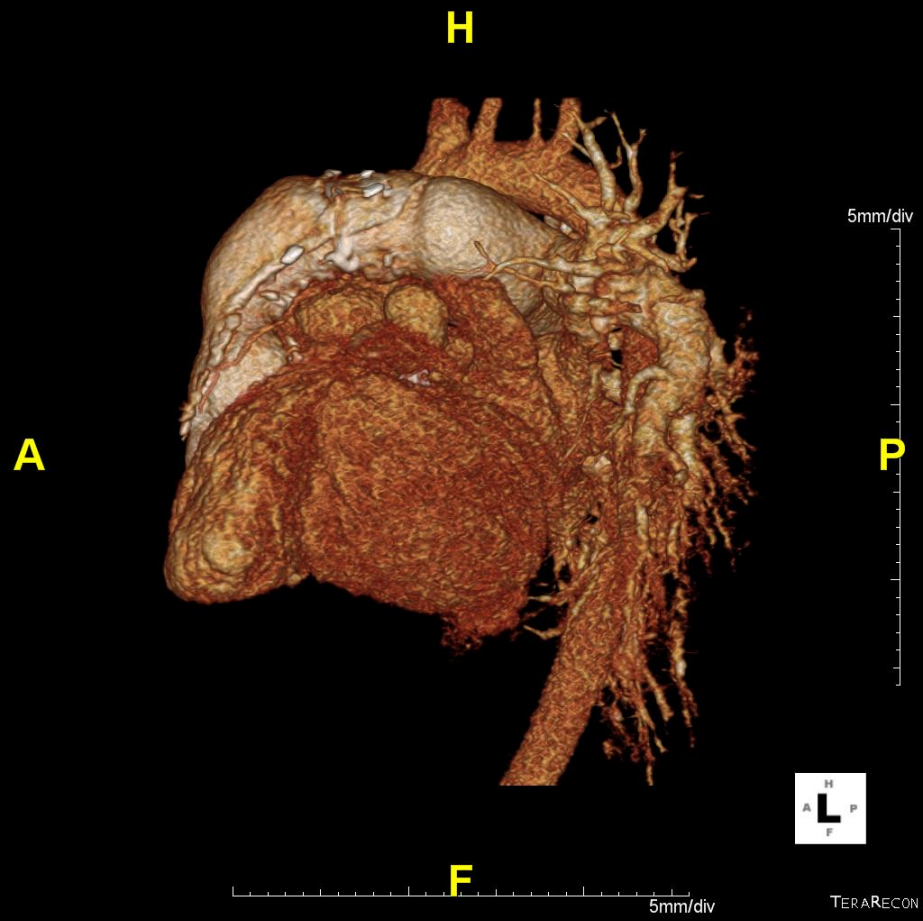
★RV/Ao =100/22 (152/92/115)

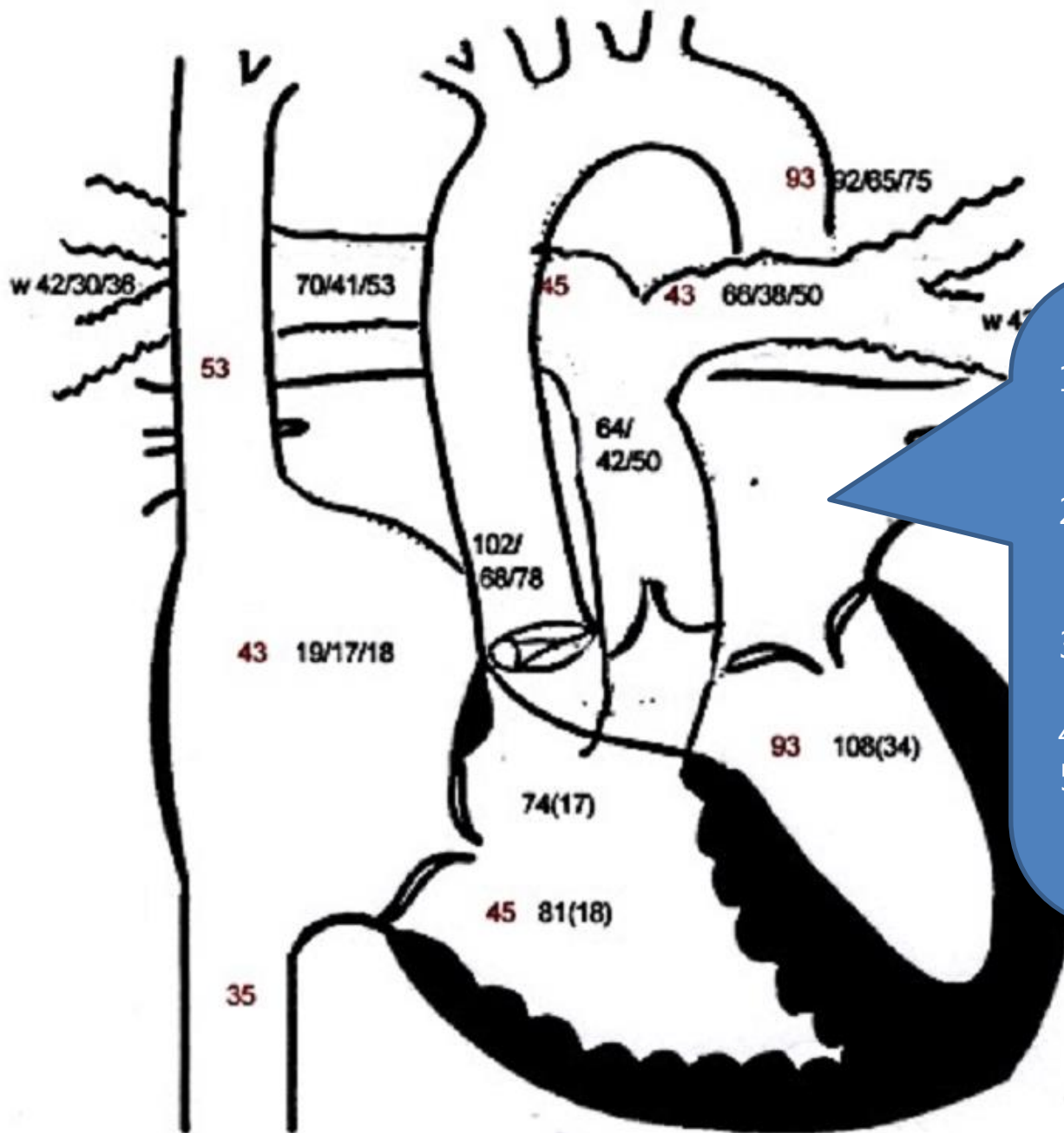
★PG=RV-RVO=70mmHg

PG(RVO-MPA)=4 mmHg

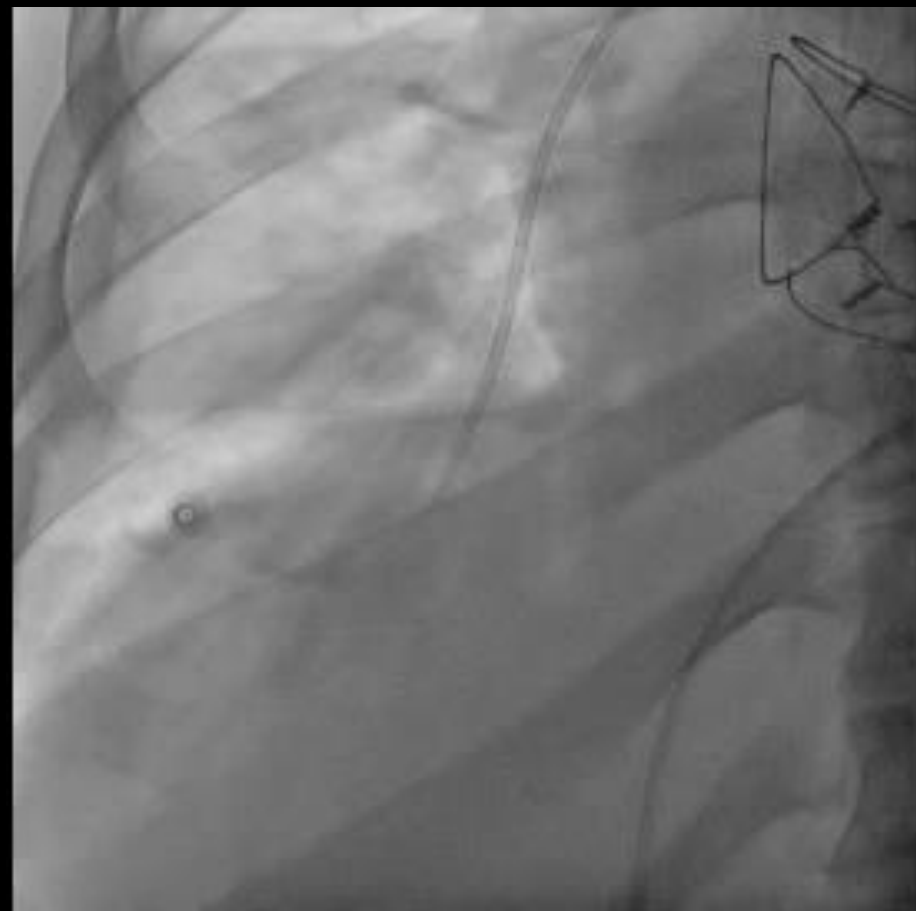


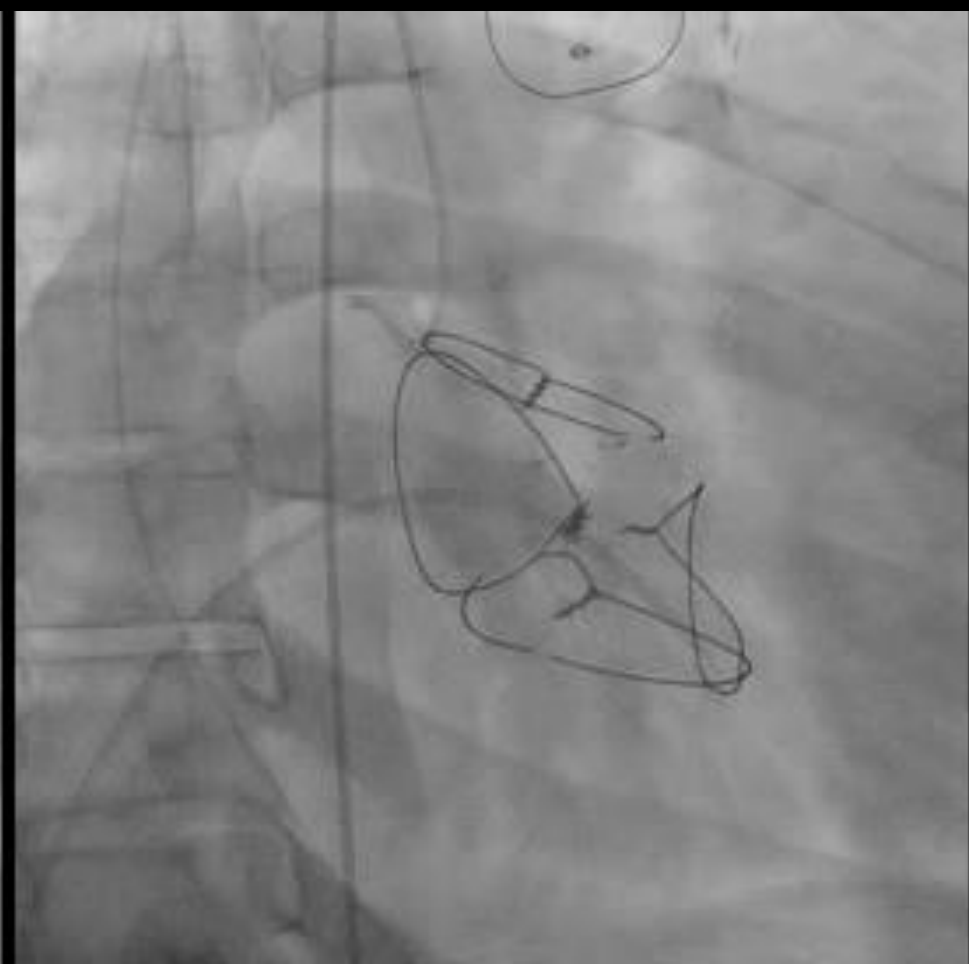






1. Severe heart failure with very high LVEDP=31~34mmHg
2. severe Pulmonary HTN mean PAP=50~53mmHg Rp=9.4 WU
3. both pulmonary vascular bed → hypertensive change (+)
4. High RVEDP=17~18mmHg
5. LCO d/t severe heart failure (Qs=1.6)





Take Home Message

- 1. Annually routine F/U.
- 2. multi-modality & meticulous approach
Echo, CT, MR, Holter, CPET
if needed, cath. & EP
- 3. collaboration with colleague