Surgical Treatments for Severe Pulmonary Regurgitation in Repaired TOF

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Surgical management of TOF

- First open heart repair by Lillehei and Varco in 1954
- Primary repair during infancy
- Trans-annular patch
- Limited RVtomy
- Trans-atrial repair

TOF Repair Survivals

- Early mortality – less than 2-3%
- TOF repair survivals

  ; 20-year survival rate (treated until 1970s)

  -90% (Murphy JG, N Engl J Med 329:593-599, 1993)
- Third postoperative decade and afterward

  ; arrhythmias, exercise intolerance, heart failure, death

Anatomic and Functional Abnormalities in Repaired TOF

- RV volume overload from PR
- Residual atrial and/or ventricular septal defect
- Tricuspid regurgitation
- RV outflow tract patch aneurysm
- Pulmonary artery stenosis

Severe Chronic Pulmonary Regurgitation

- Important, treatable cause of RV dilatation and failure
- Pulmonary valve replacement
  ; indications
  methods
  optimal timing
- Pathophysiology and its management

Isolated Congenital PR

- 72 cases
- At age 20 years - 6% symptoms
  At age 40 years – 29% symptoms
- Symptomatic three pts – death followed after the appearance of sx(39 Months)
- Probability of heart failure – increased exponentially after age 40 years

(Shimazaki Y, Thorac Cardiovasc Surg 32:257-259,1984)

Factors Determining the Degree of PR

- Size of the regurgitant orifice
- Diastolic pressure gradient between the PA and RV
- Capacitance of the PAs
- Duration of diastole

- Severity of PR ; increase over time

(Kuehne T, Circulation 108:2007-2013,2003)

PR at the Time of TOF Repair

- RV –hypertrophied, low compliance
- Central PA –hypoplastic, low capacitance
- Short duration of diastole

Limit the degree of PR

(Geva T, Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 9:11-22,2006)

PR long after TOF Repair

- RV stroke volume ↑
- Size and compliance of central PAs ↑
- RV compliance ↑
- Longer duration of diastole

Increase in the degree of PR over time

(Geva T, Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 9:11-22,2006)

Response of LV to Severe Chronic AR

I Compensated stage; end-diastolic volume ↑, chamber compliance ↑, normal mass-to-volume ratio (compensated hypertrophy)

II Failure of compensatory mechanisms; mass-to-volume ratio ↓, end-systolic stress ↑, global systolic function ↓

III Further LV dilatation; Progressive global systolic dysfunction

IV Irreversible myocardial injury; fibrosis

(Chaliki HP, Circulation 106:2687-2693,2002)

PR is different from AR

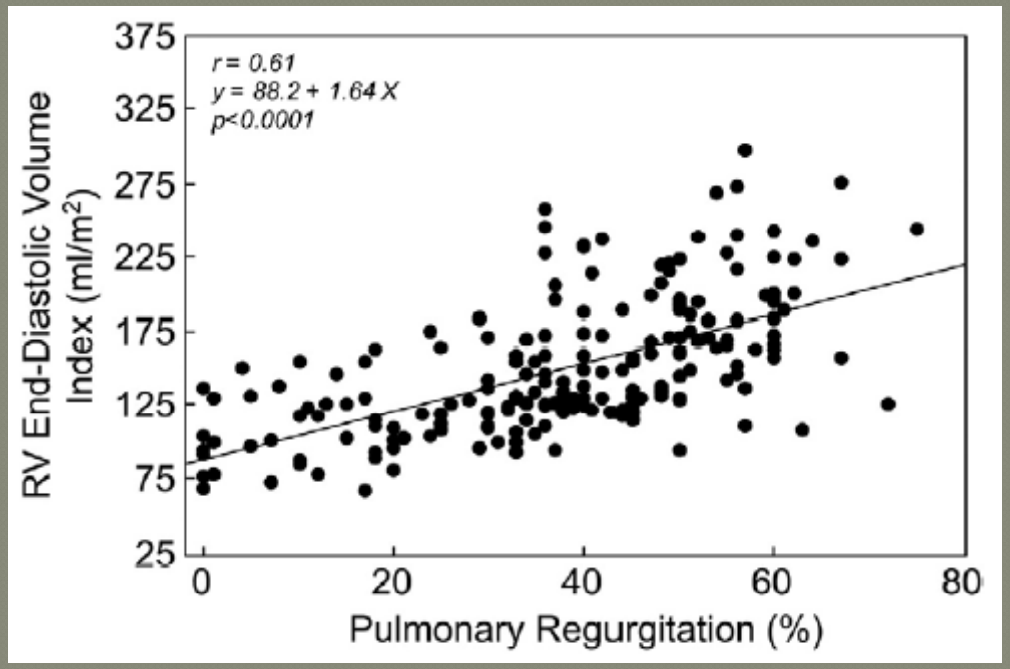
- RV – temporarily acts as a conduit during late diastole(late diastolic forward flow in the PA)
- Pulmonary microvascular bed – low resistance , located close to the heart

→ valve-like effect in severe PR

Severe PR – only 40% regurgitant fraction, well tolerated for a long time

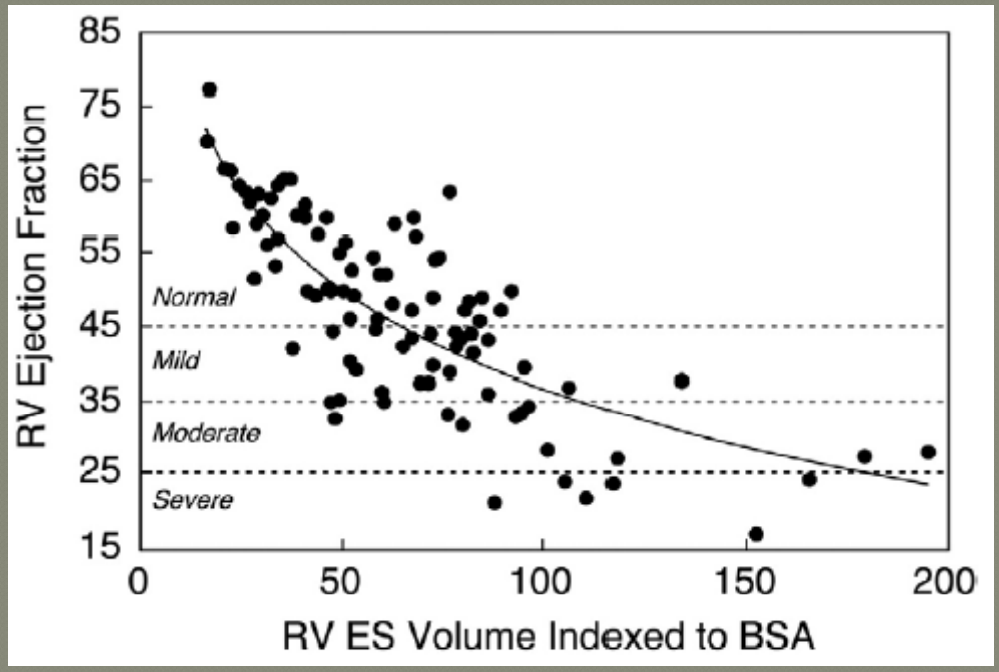
(Bouzas B, Europian Heart Journal 26:433-439,2005)

RV Mechanics After TOF Repair



(Adapted from Geva T, Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 9:11-22,2006)

RV Mechanics After TOF Repair



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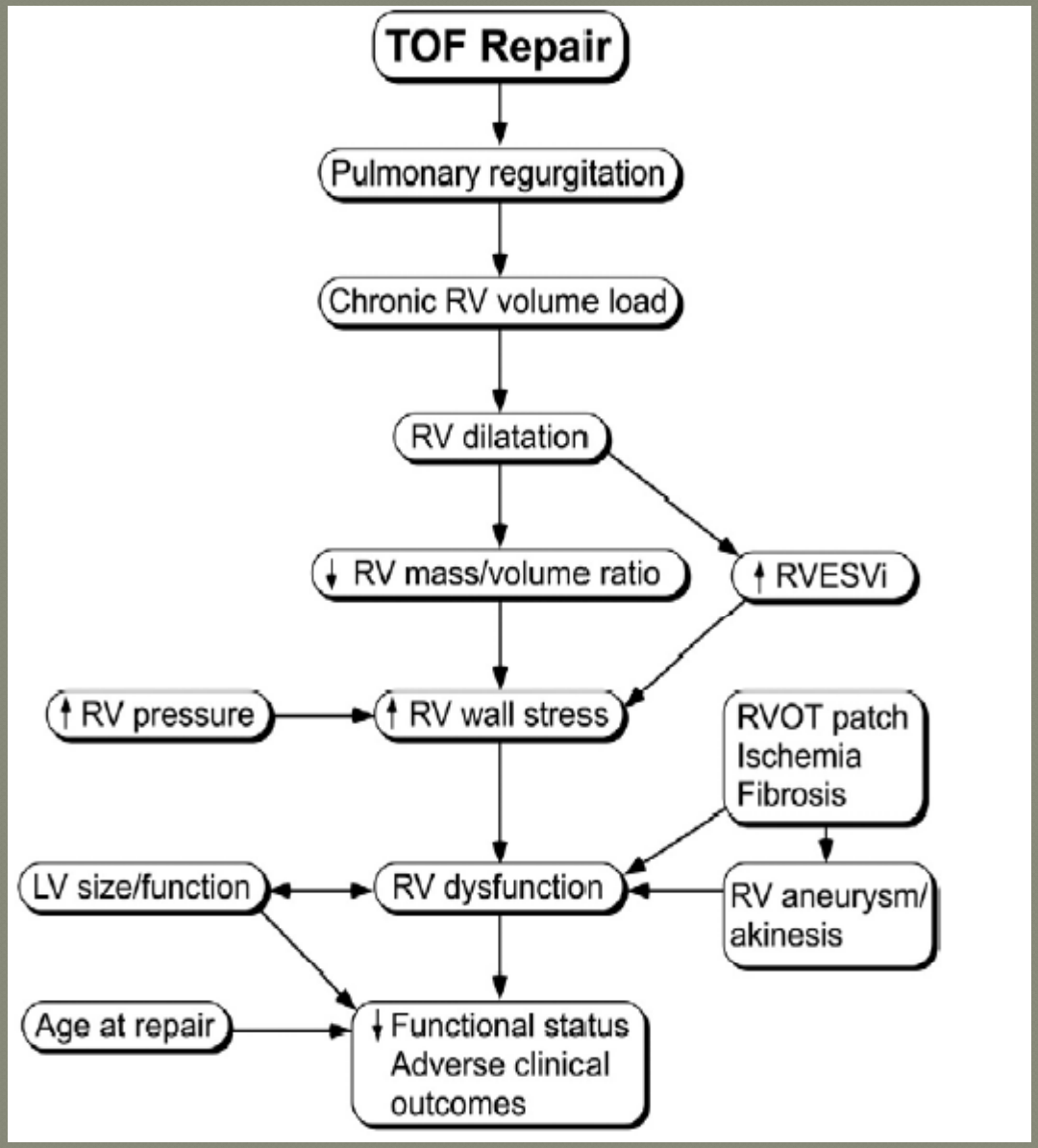


(Adapted from Geva T, Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 9:11-22,2006)

Clinical Course and Outcomes After TOF Repair

- Exercise intolerance
- Heart failure
- Arrhythmias
- Death

  ; The mortality rate nearly triples during the third postoperative decade

(Murphy JG, N Engl J Med 329:593-599, 1993)

Exercise Intolerance I

- More than 10 yrs after repair ; 80-90% predicted
- Cardiomegaly, RV hypertension, pulmonary regurgitation, residual VSD, pulmonary H/T, arrhythmias
- Restrictive lung defect in repaired TOF ; diminished lung volumes related to cardiomegaly, PR

(Wessel HU, J Thorac Cardiovasc Surg 80:582-593,1980)

Exercise Intolerance II

- Mean exercise duration ↓,Peak oxygen consumption ↓,Vital capacity ↓
- Benefit of a stiff RV in chronic PR
  ; smaller CT ratio, higher peak oxygen consumption (Gatzoulis MA, Circulation 91:1775-1781,1995)
- The effect of PR on LV function; RV dilatation may affect LV performance
  ; Cardiac output and ventriculography measured during cycle ergometry
  (Kondo C, Circulation 92:250-255,1995)

Arrhythmias and Sudden Death I

- QRS duration≥180 ms (strongly predictive)
  ; ventricular tachycardia, sudden death, atrial flutter/fibrillation
- Previous Water or Potts shunt (modest effect)
  ; ventricular tachycardia
- Older age at time of repair; sudden death

(Gatzoulis MA, Lancet 356:975-981,2000)
;Multicenter study of 793 pts, f/u of 21 yrs

Arrhythmias and Sudden Death II

- Transatrial-transpulmonary repair
  → less RV dysfunction,
  fewer arrhythmias (w/o RVtomy)
  less severe PR

(Dietl CA, Circulation 90(suppl II)7-12,1994)

- Combined LV and RV dysfunction
  → Arrhythmia risk ↑
  (ventricular interdependence)

(Ghai A, J Am Coll Cardiol 40:1675-1680,2002)

Survival

- Transannular patch
  - predictor of early postoperative death
  - little impact on late survival

(Kirklin JK, Ann Thorac Surg 48:783-791,1989)

- More recent report
  - transannular patch(PR)
    : considerable impact on long-term outcome

(Murphy JG, N Engl J Med 329:593-599,1993)

- Cardiac deaths
  - sudden death or heart failure

Methods of Investigation I

- Hx ; age at TOF repair, time from repair, syncope, Sx of heart failure
- ECG ; QRS duration, QRS change
- Echocardiography

  ; main limitation of image quality , accuracy of measurements of RV volumes and PR

  →Harmonic and contrast imaging tech, Tissue Doppler imaging

Methods of Investigation II

- CMR ; accurate quantitative information on biventricualr size, function and blood flow measurements
- CT ; ECG gating CT →reconstruction of temporally resolved images of the ventricles and calculation of volumes and EF
- Exercise test
  ; objective assessment of exercise capacity, exercise induced arrhythmia
- Cardiac catheterization

Indications and Timing of PVR

- Severe PR with Sx →PVR
- Severe PR w/o Sx →prophylactic PVR?
- Risk/Benefit analysis
  - Natural history
  - Pathophysiology
  - Risk factors for adverse outcomes if no intervention
  - Risk of the procedure
  - Its potential benefits

Predictors of Adverse Clinical Outcomes

- Moderate-severe RV dilatation
- RV dysfunction
- LV dysfunction
- QRS duration ≥180 ms
- Syncope
- Positive programmed ventricular stimulation study
- Sustained ventricular tachycardia

Risks of PVR

- Operative mortality ; low(1-2%)
- Continued low risk of death after PVR
- Survival after PVR ;

  92% at 5 Yrs, 86% at 10 Yrs

  (Therrien J, Circulation 103:2489-2494,2001)

  95% at 5 Yrs, 76% at 10 Yrs

  (Discigil B, JTCS 121:344-351,2001)
- Freedom from reoperation ;

  81% at 5 Yrs, 58% at 10 Yrs, 41% at 15 Yrs

  (Calderone CA, JTCS 120:1022-1030,2000)

Benefits of PVR

- Symptomatic improvement
- RV end-diastolic and end-systolic volumes ↓
- RV E/F ; no significant change
- Improved exercise tolerance
- Ventricular tachycardia incidence ↓
- No clear evidence of survival improvement

Wrong RV Mechanics after PVR

- Preop EDV index≥170mL/m^2
  ; RV size did not return to normal
  (Therrien J, Am J Cardiol 95:779-782,2005)
- Severe RV dilatation and moderate dysfx
  ; no improvement in RV size and Fx
  (Therrien J, J Am Coll Cardiol 36:1670-1675,2000)

Criteria for PVR (Adapted from Geva T, Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 9:11-22,2006)

1. RV EDV index ≥ 160 mL/m^2
2. RV ESV index ≥ 70 mL/m^2
3. LV EDV index ≤ 65 mL/m^2
4. RV EF ≤ 45%
5. RVOT aneurysm
6. Clinical criteria
   : exercise intolerance, heart failure, cardiac medications, syncope, sustained ventricular tachycardia

- Other hemodynamically significant lesions
  ; case by case
- TOF repair at age ≥3 yrs ; sooner

Surgical Options for PVR I

- Mechanical prostheses, heterografts, homografts, autologous pericardial valves, bovine jugular valves mounted on stents

- Mechanical valve
  ; frequent thromboembolism and valve failure(low-pressure, slow-flow)
- Single-tilting disc valve
  ; low opening pressure,
  lack of hinge mechanisms

(Rosti L, Ann Thorac Surg 65:889-890, 1998)

Surgical Options for PVR II

- Valves of choice for PVR
  - Bioprosthetic valves(homograft or heterograft)
    - avoidance of anticoagulation
    - accelerated calcification in children
    - long life span in adults(15-30 yrs)
  - Homograft
    - excellent hemodynamic performance
    - availability ↓, size limit
    - early failure by immunologic basis

(Welters MJ, Hum Immunol 63:1019-1025, 2002)

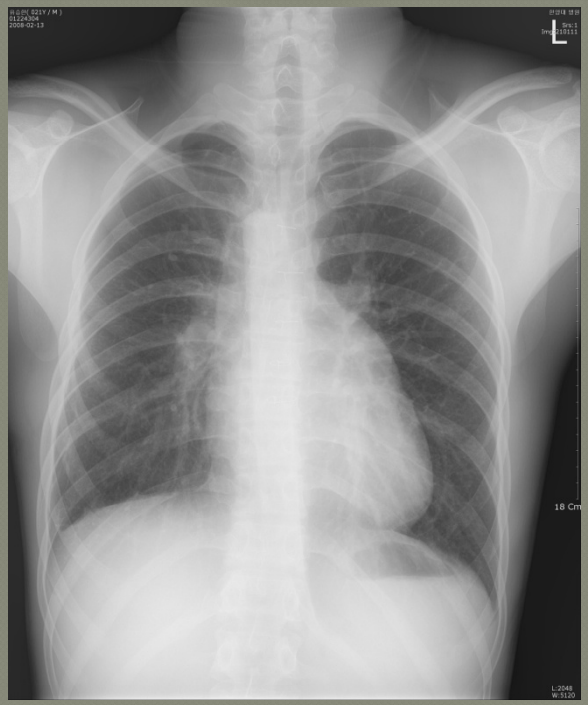
Surgical Options for PVR III

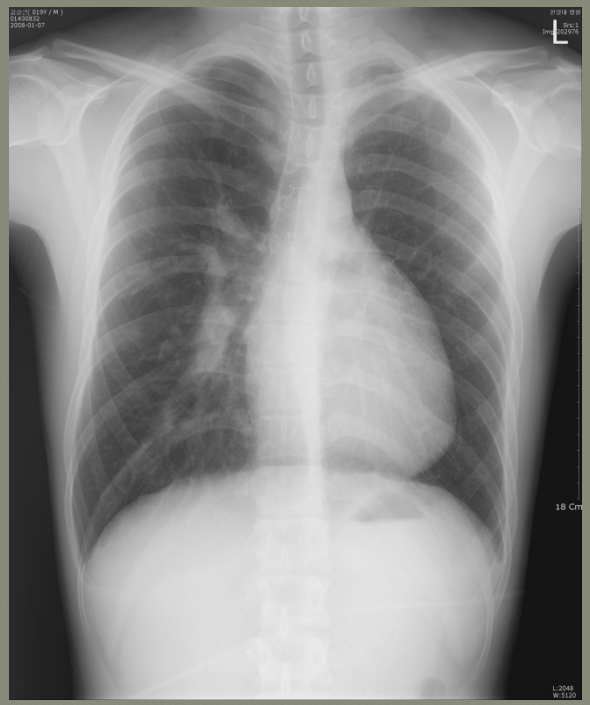
- Stentless bioprosthetic valve
  ; Dissipation of mechanical stress on the valve leaflet
  → structural failure ↓, durability ↑
  (Park SZ, Curr Opin Cardiol 15:74-81, 2000)
  ; Effective valve orifice size 2-4 mm larger than stented
  (Reardon MJ, Curr Opin Cardiol, 14:84-89, 1999)

Conclusion

- Severe PR in repaired TOF
  - not a benign lesion
  - detrimental hemodynamic & clinical effect
- PVR should be performed before irreversible RV dysfunction
- Ideal prosthetic valve for RVOT reconstruction
- Markers of the reversibility of ventricular dilatation; elusive

Clinical Cases





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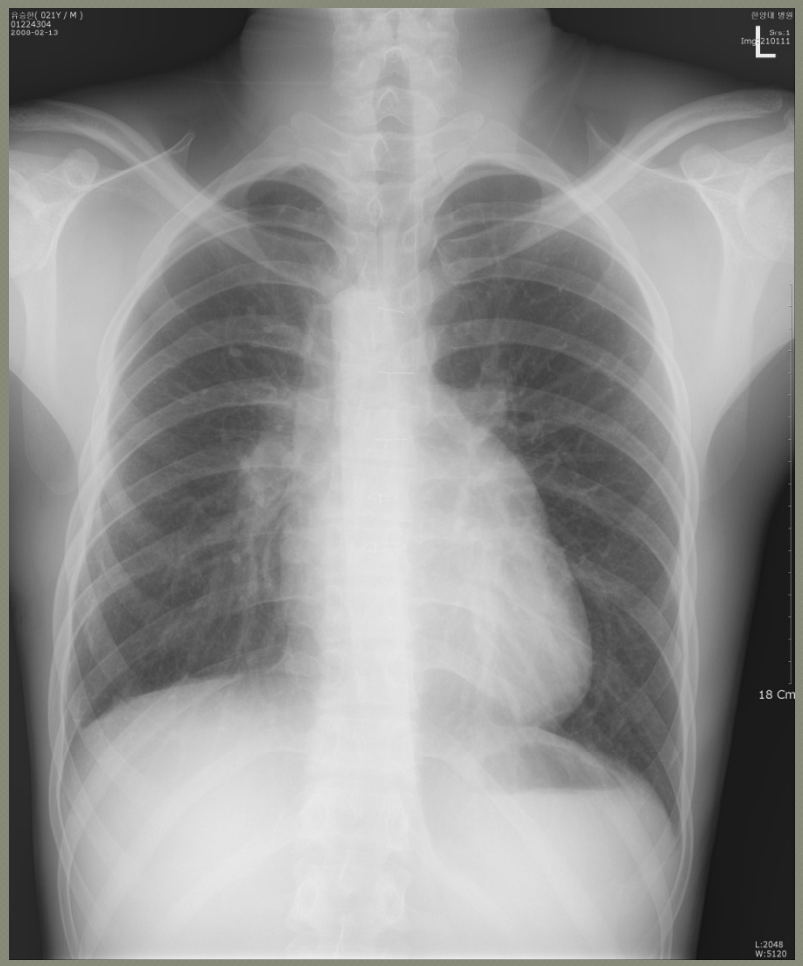
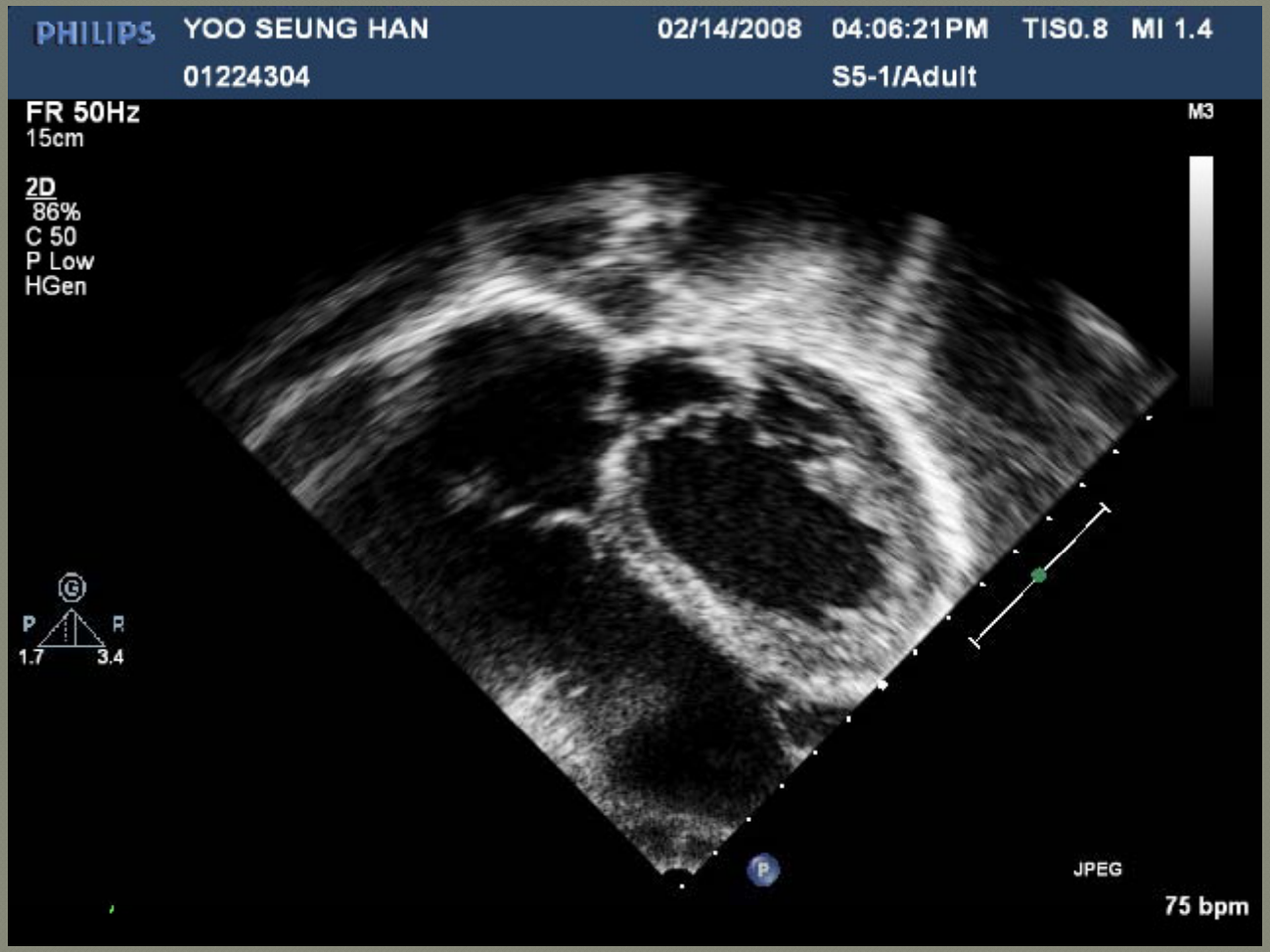


- Modified BT shunt
  - 3 mo old
- Total correction
  - 2 yrs old
- Duration
  - 19 yrs
- QRS: 190ms

Case I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75 bpm

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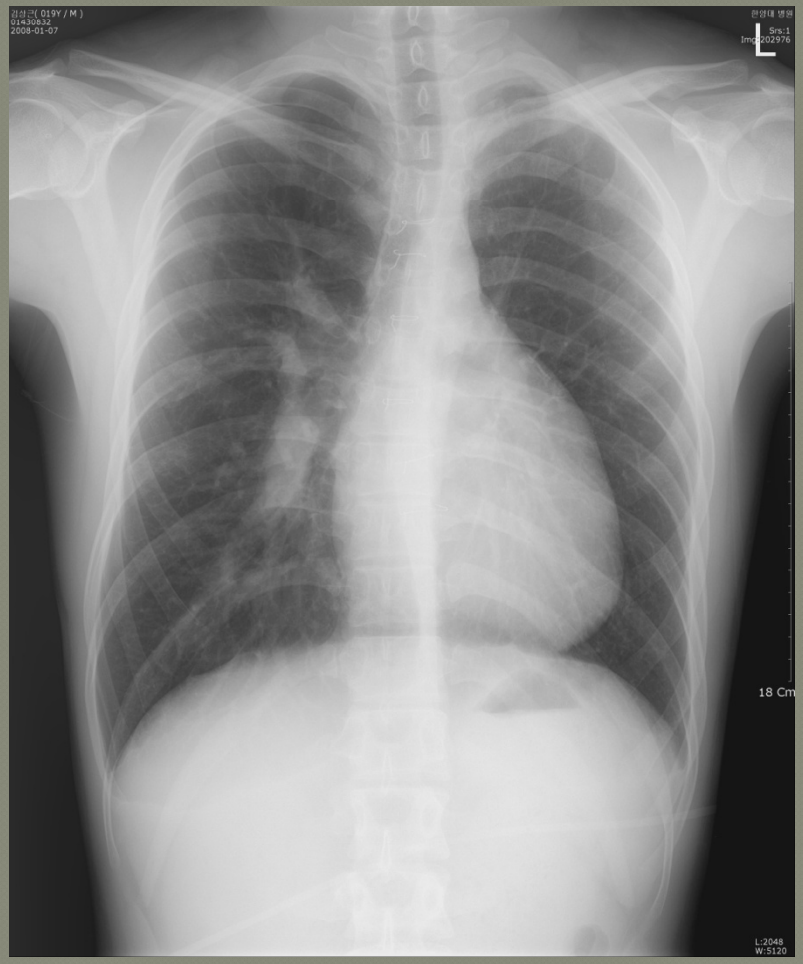
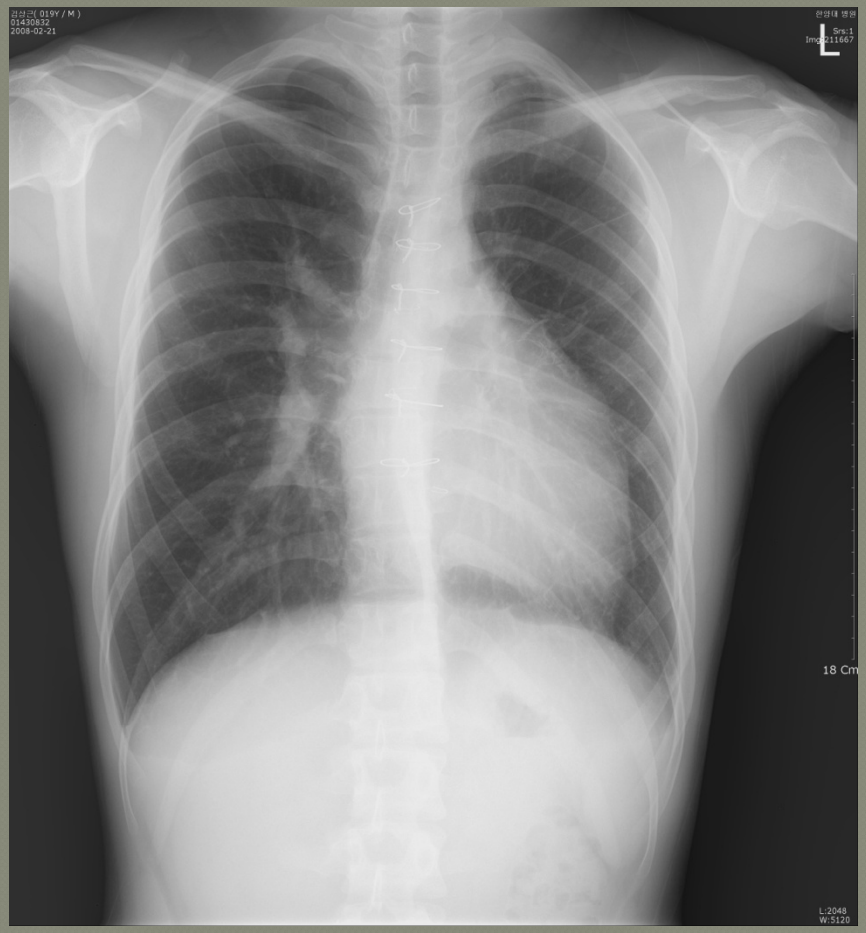
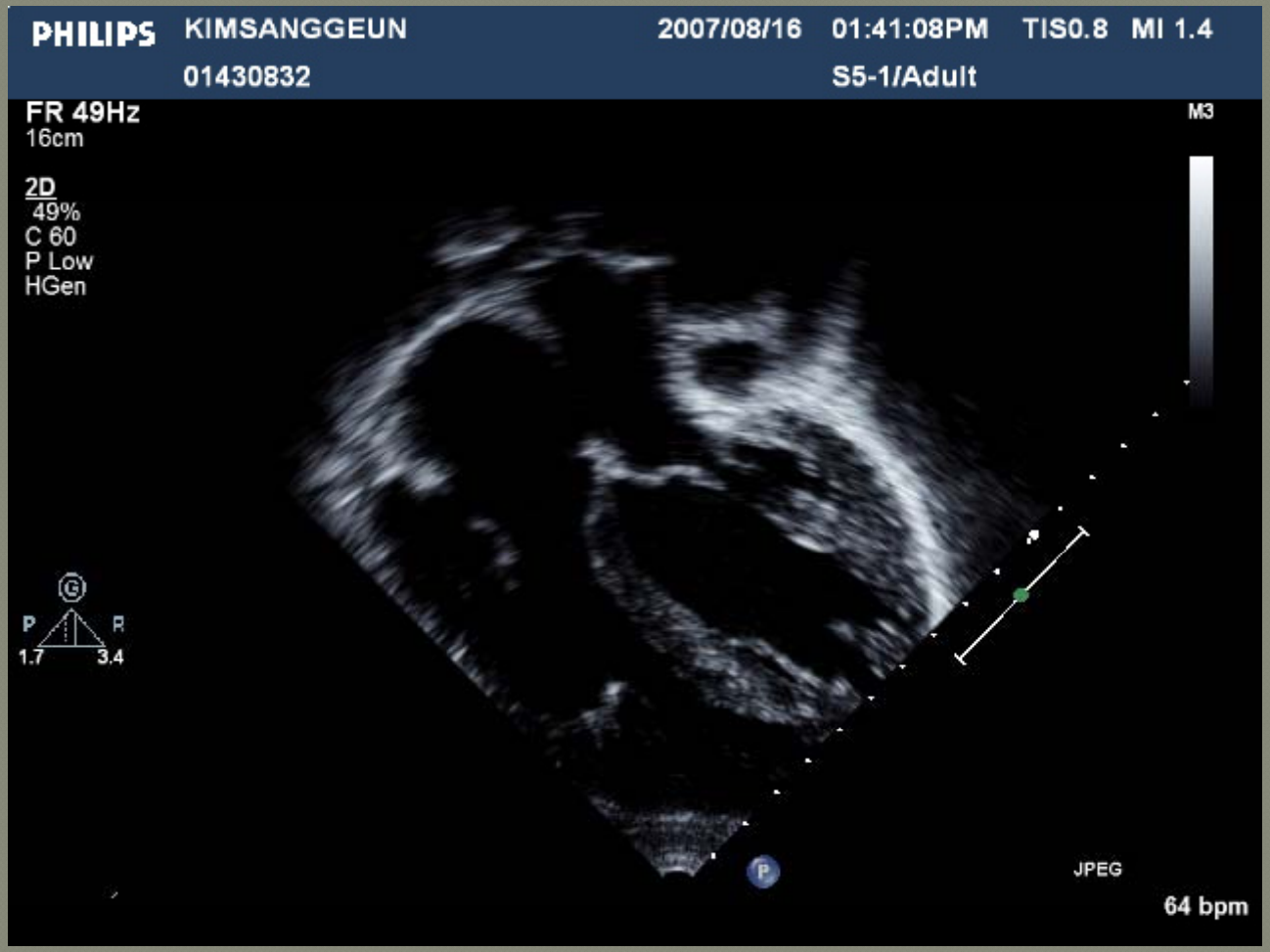




Case II (M/19)

- Total correction
  - 1 yr old
- Duration
  - 18 yrs
- Exercise intolerance
- QRS: 190 ms
- PVR with Epic 25mm, LPA angioplasty, ASD closure, TV annuloplasty

Case II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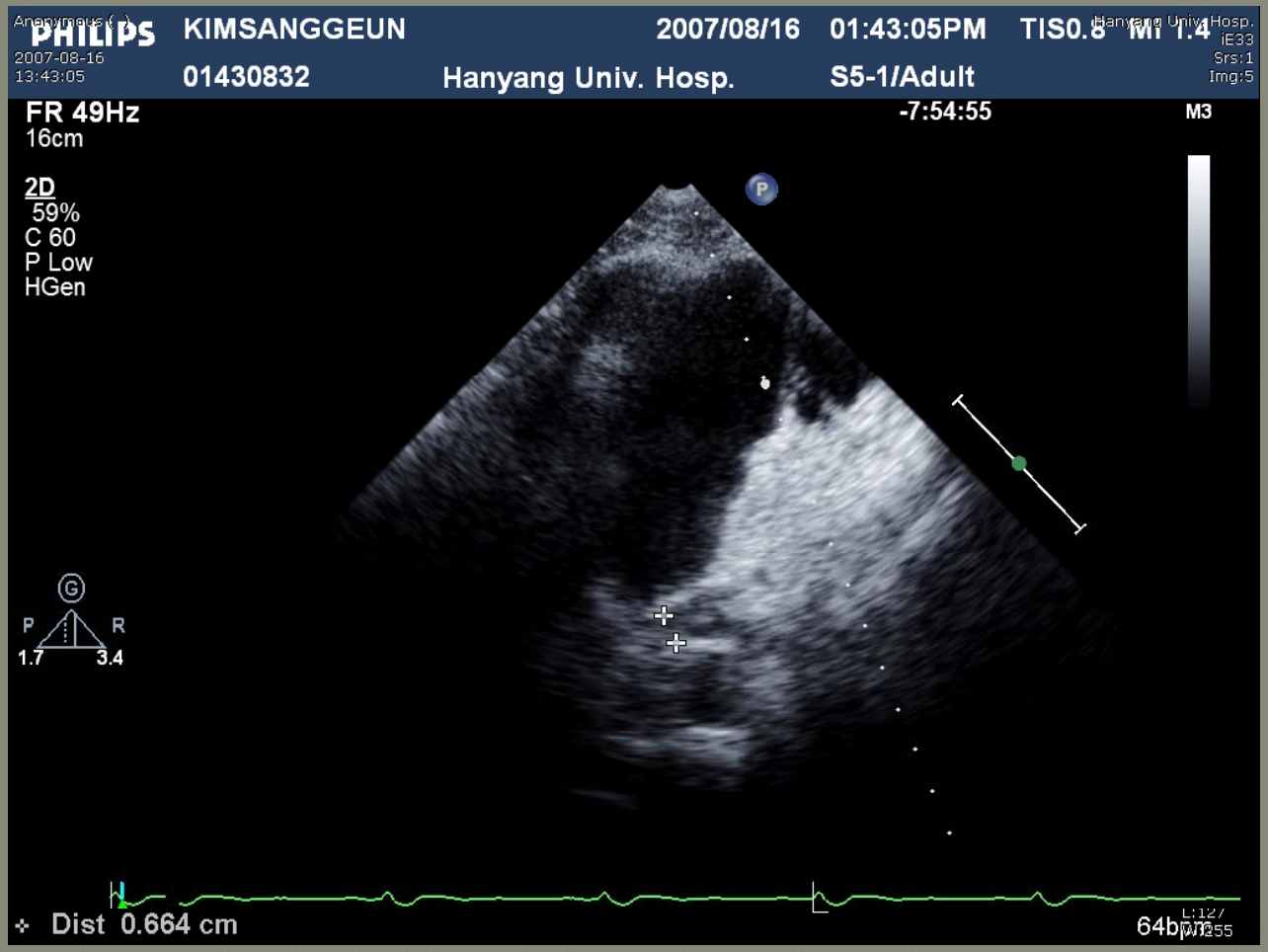
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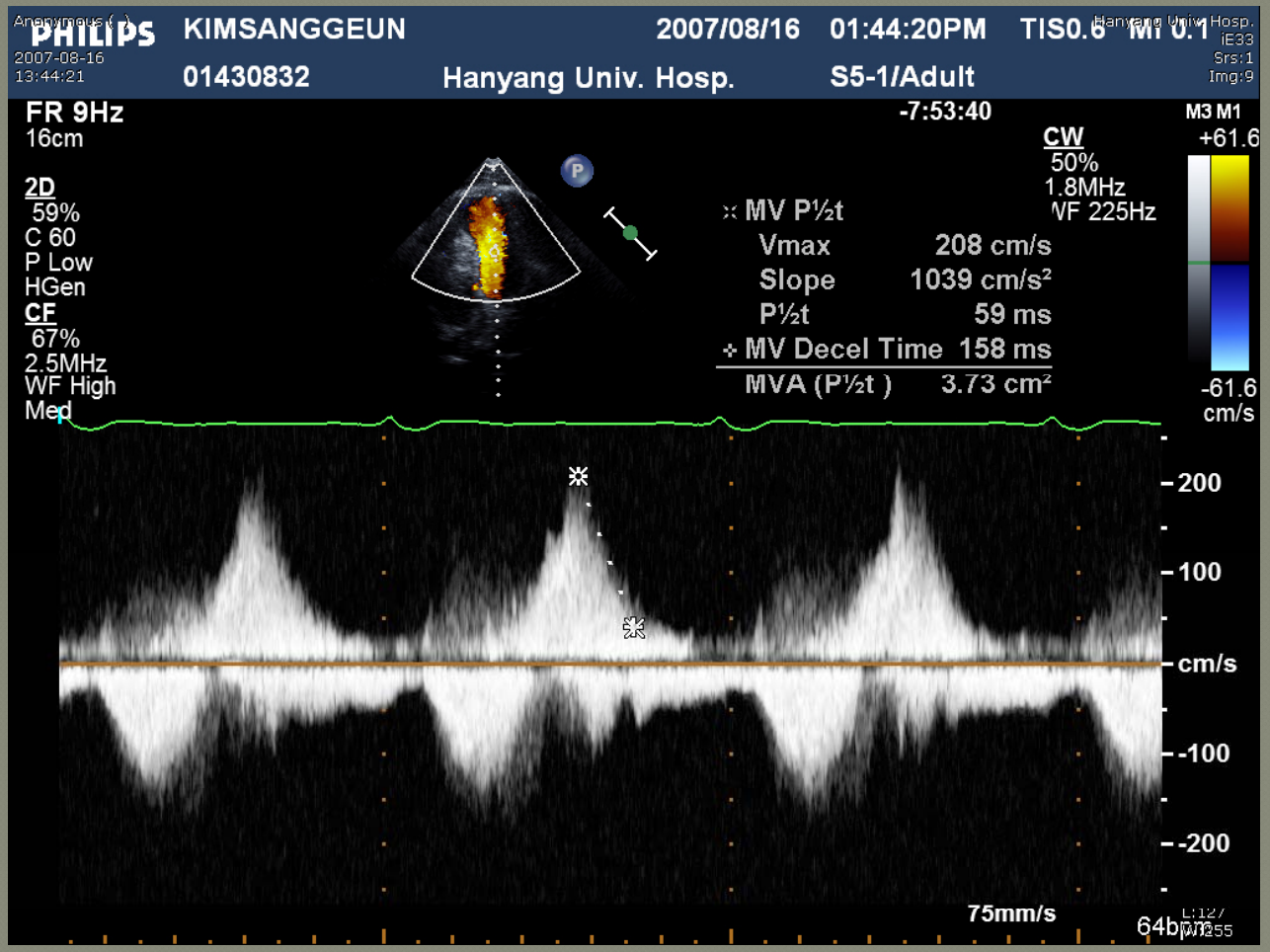
Case II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50%
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Slope 1039 cm/s²
P½t 59 ms
MV Decel Time 158 ms
MVA (P½t) 3.73 cm²
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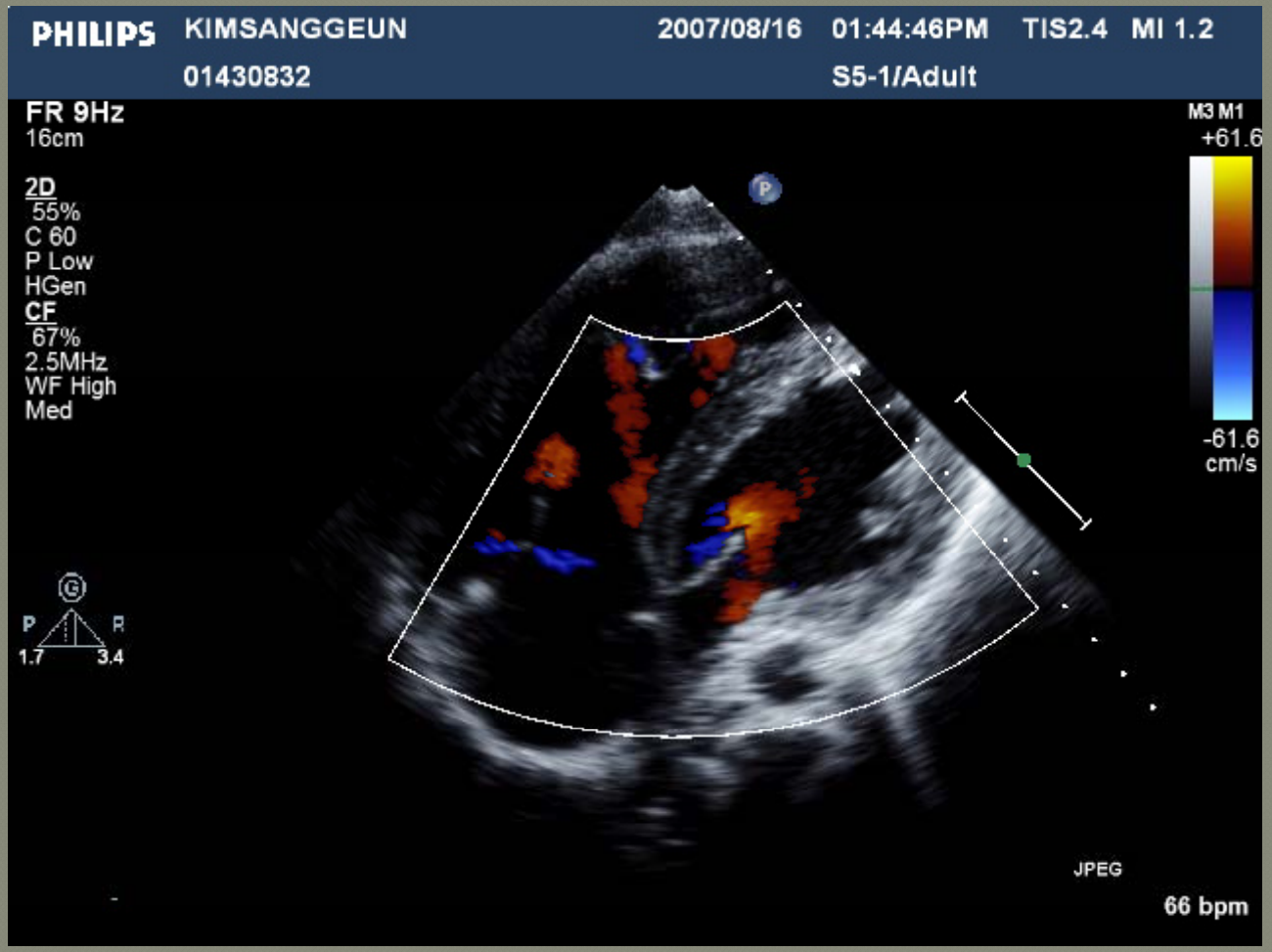
75mm/s
64bpm

Case II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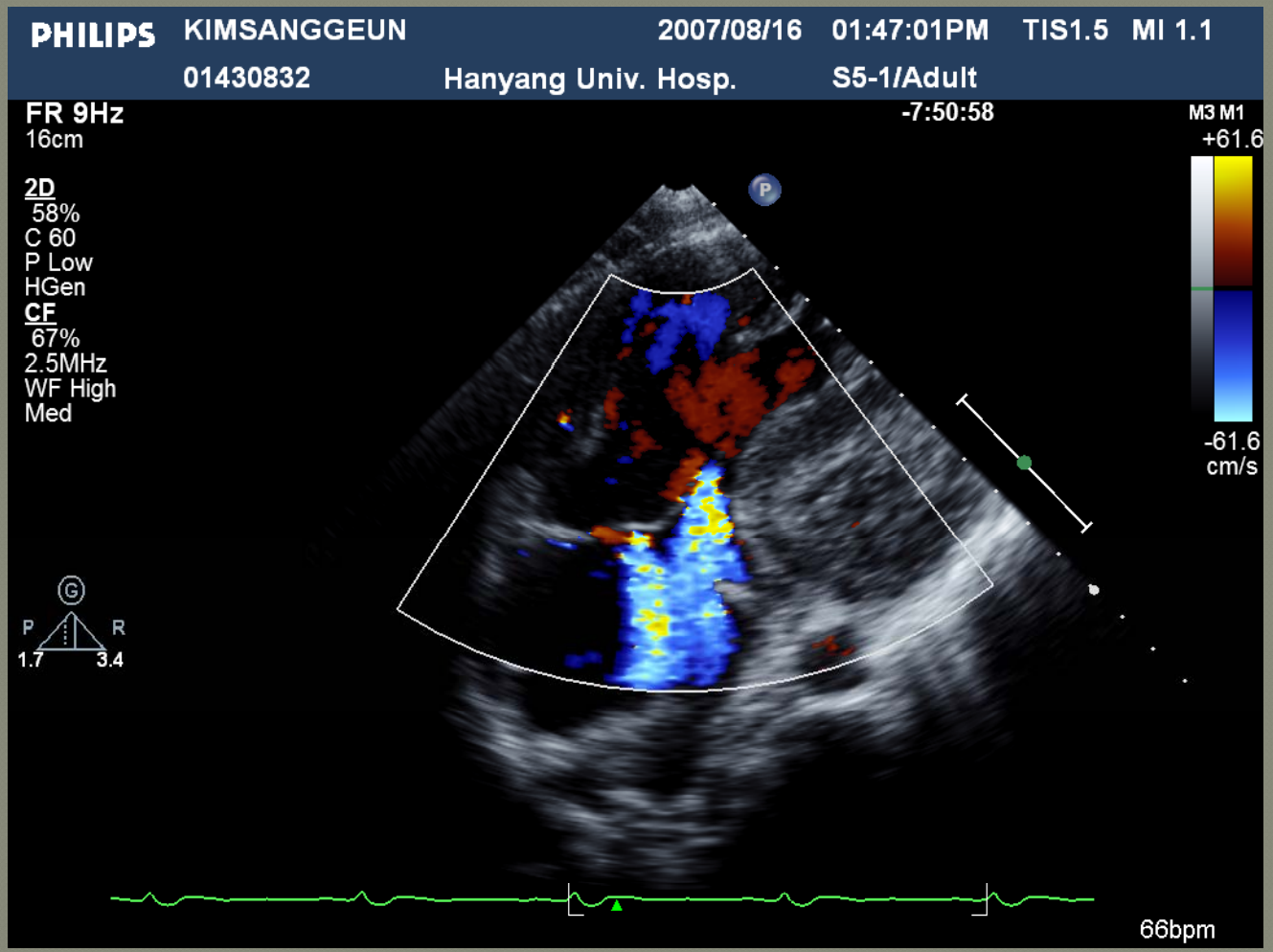
66 bpm

Case II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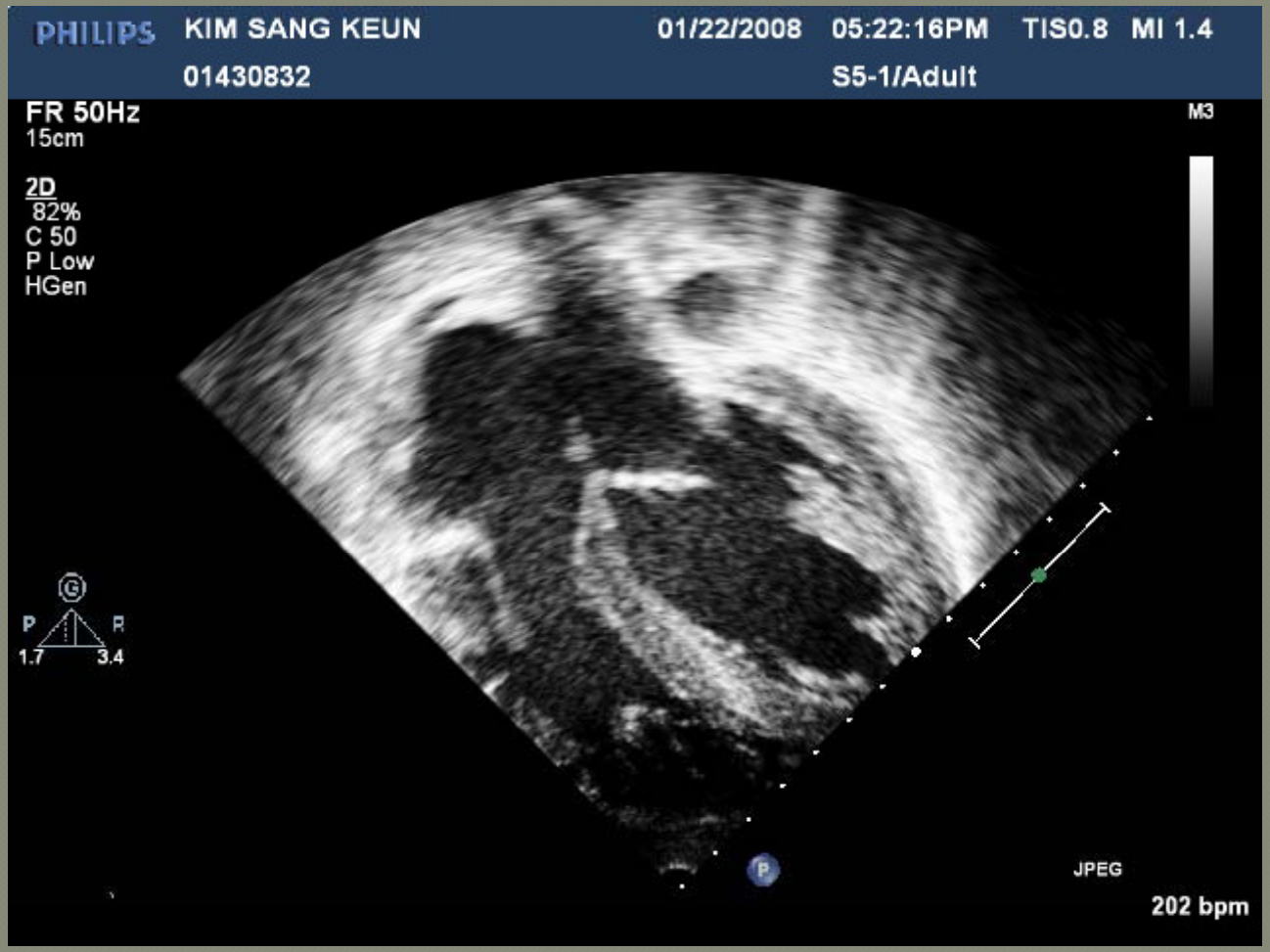
Case II

Case II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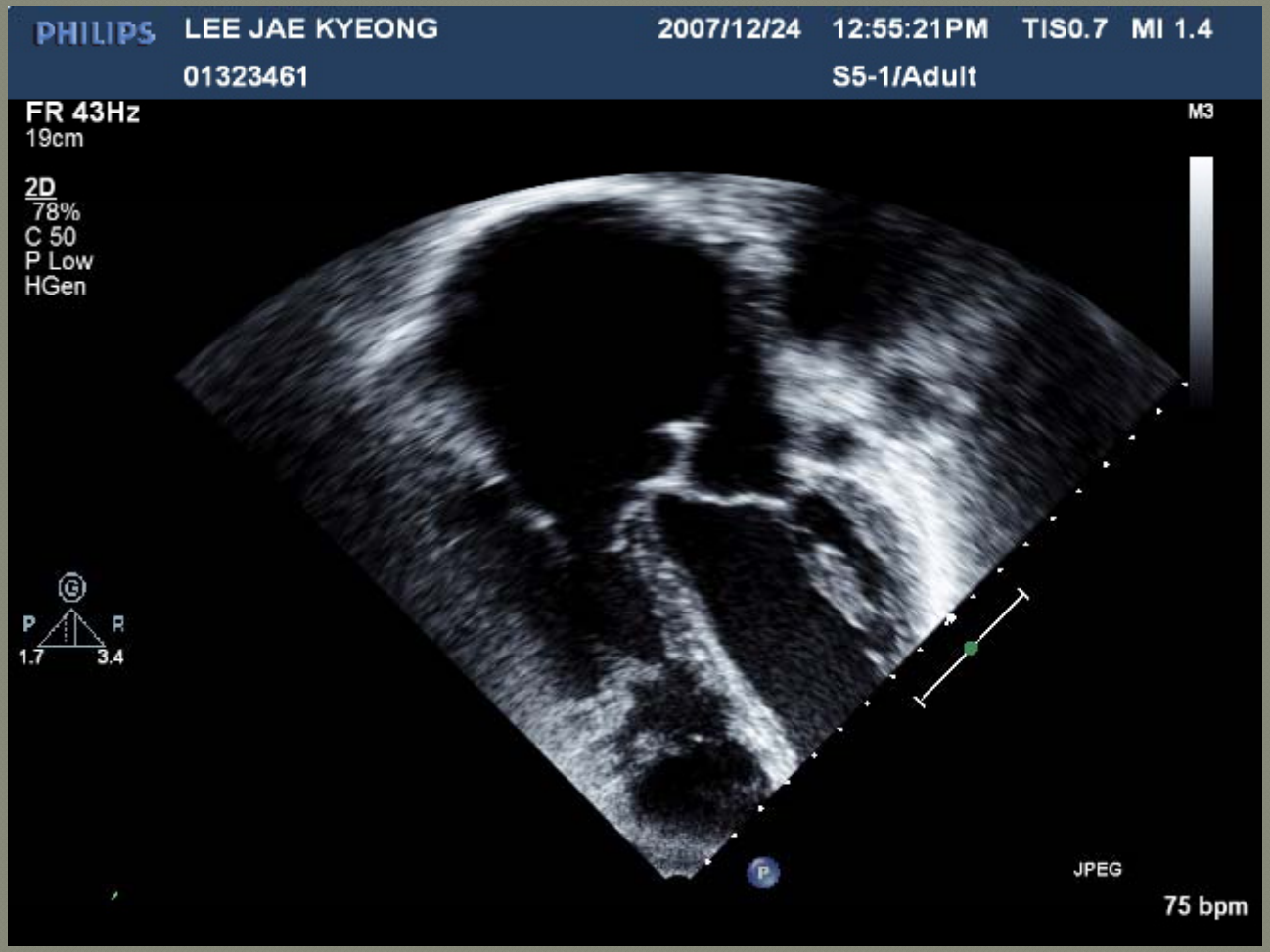
202 bpm

Case III (M/37)

Case III

- Total correction
  - 19 yrs old
- Duration
  - 18 yrs
- Exercise intolerance: Right heart failure
- QRS: 187 ms, PVCs on Holter (42/hr)
- Cardiac medications

Case III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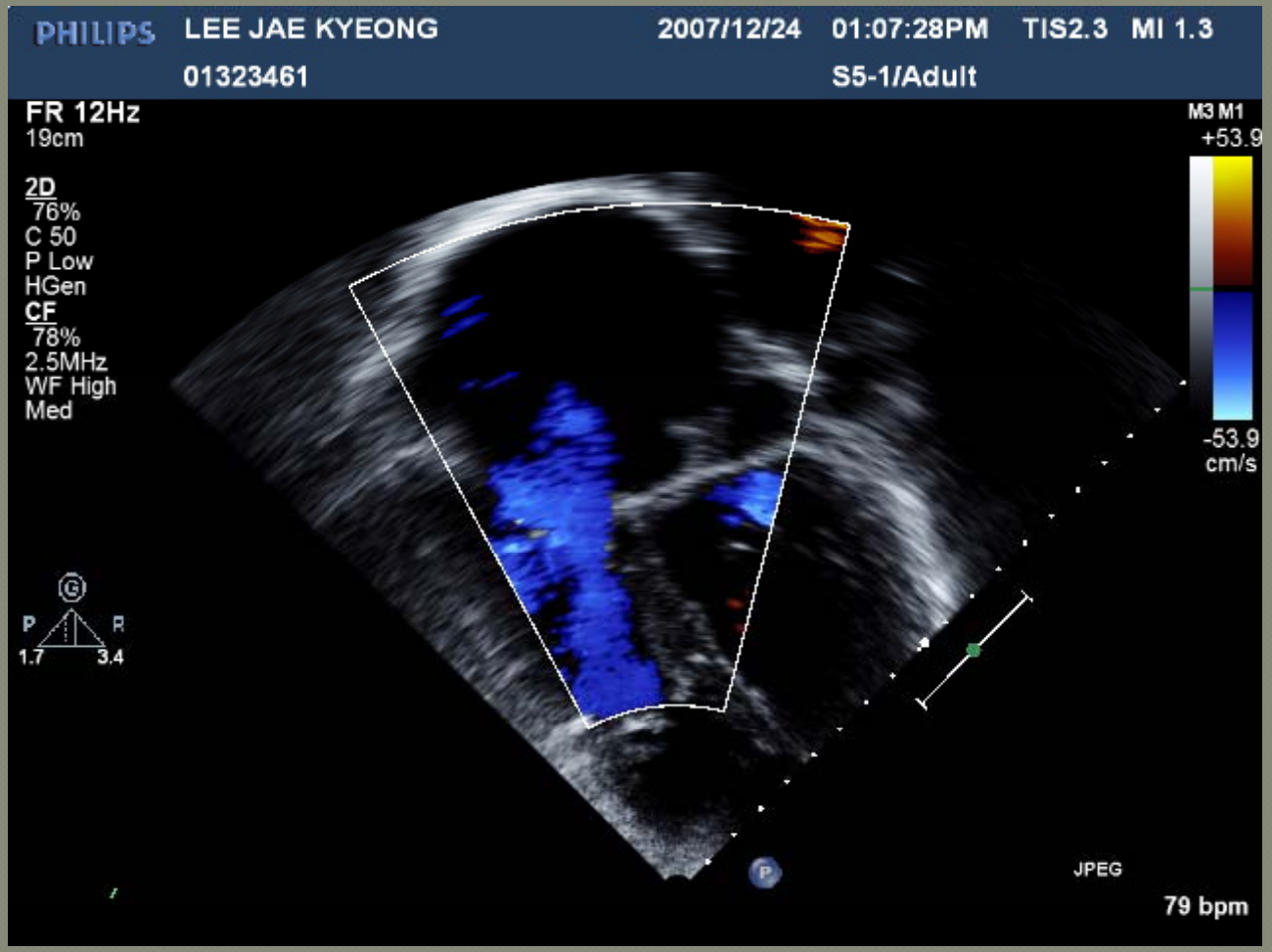
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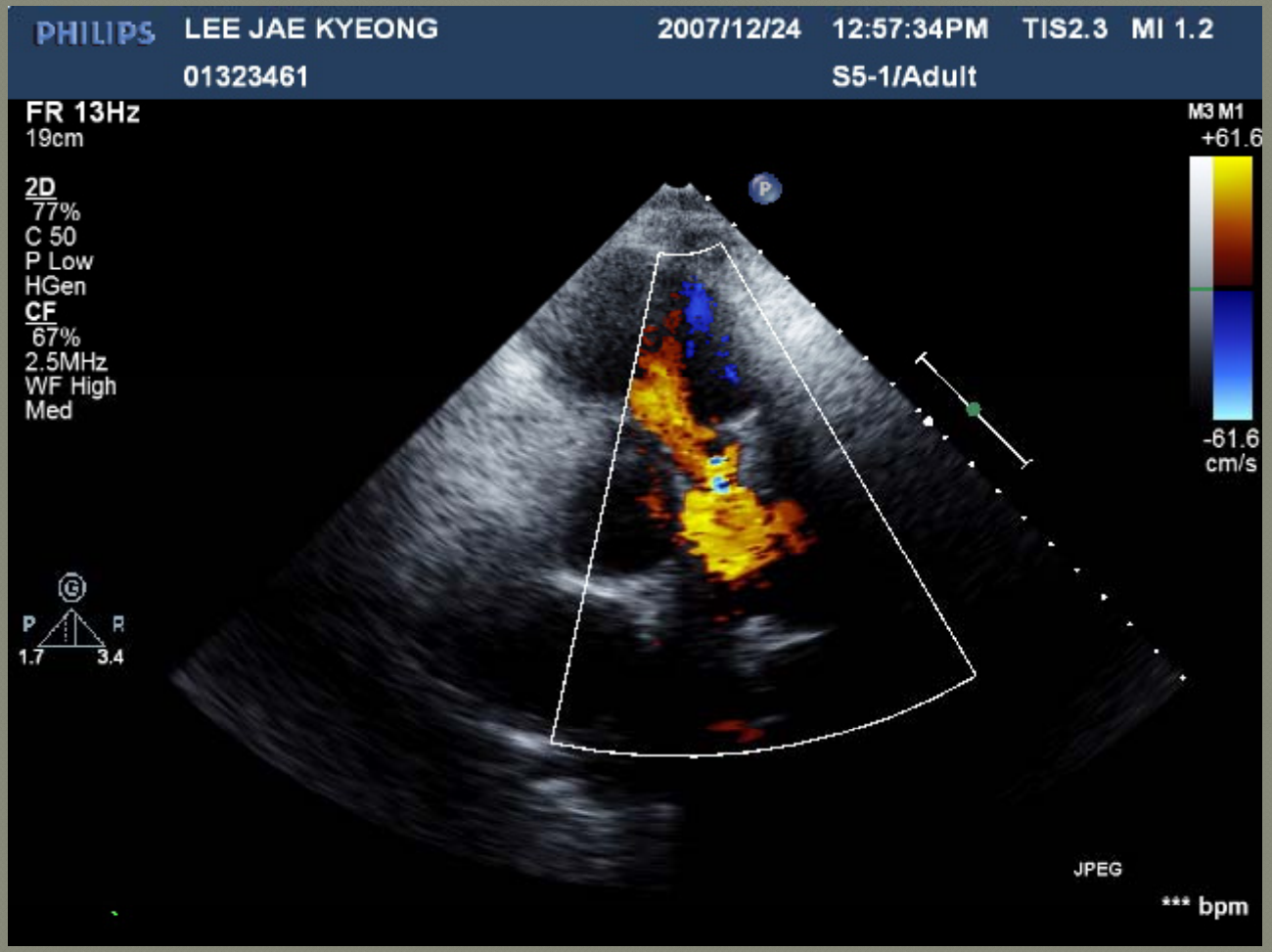
Case III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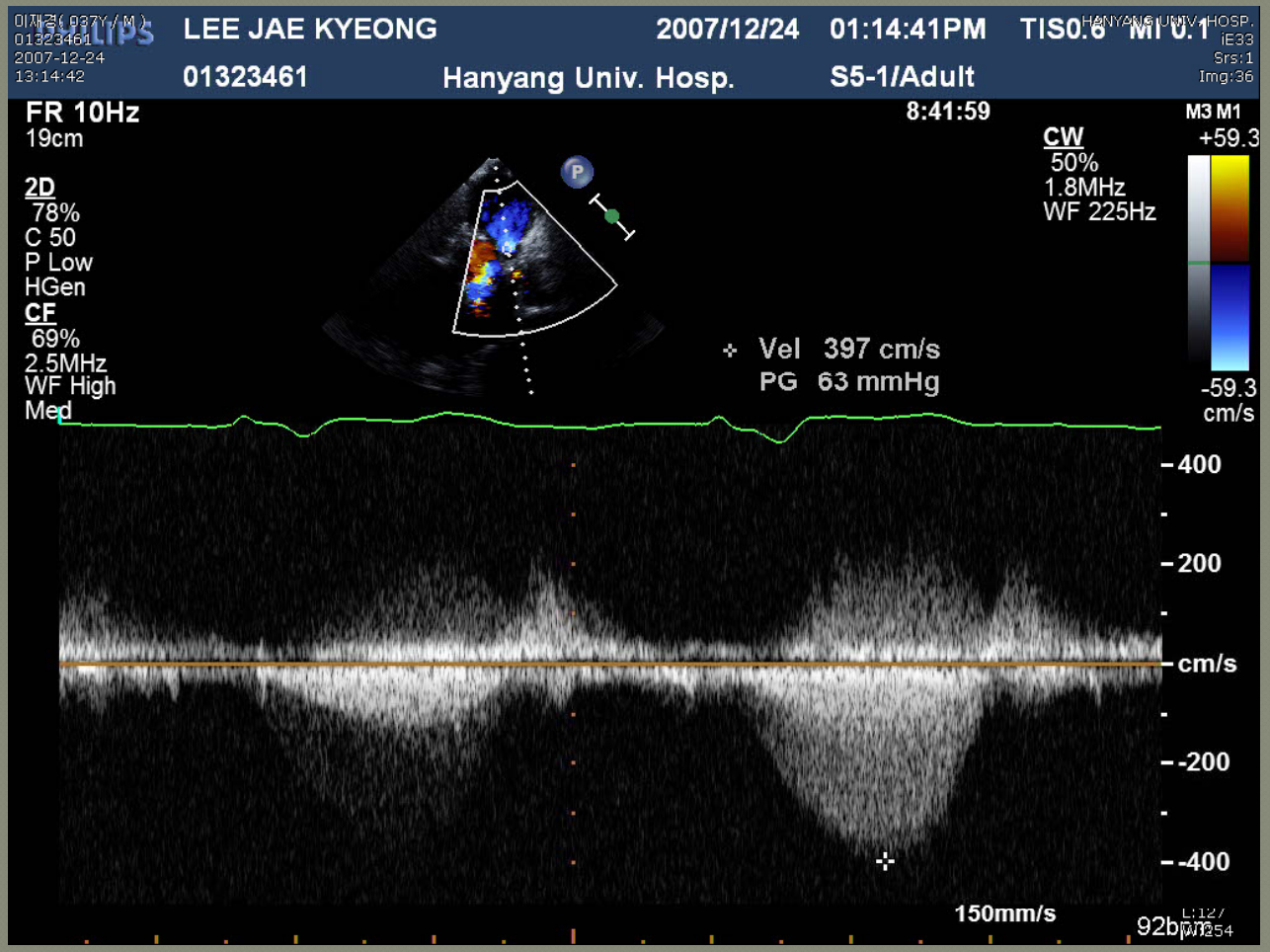
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