ACHD Case Presentation

Dr. Preeti Choudhary/Dr Rachael Cordina

Westmead and Royal Prince Alfred Hospitals, University of Sydney, Australia

Case Ms. L – double discordance and VSD

- Neonatal period –cardiac failure
- Catheterisation age 3 months - (1.5:1 shunt), PA pressures ¼ systemic
- Digoxin, diuretics
- Growth along 3rd centile
- Early tricuspid valve incompetence

Family Hx

- Female sibling died aged 5 days – complex cyanotic congenital heart disease
- Mother – died in 1980s
- Father - Type 2 DM, hypertension
- Other siblings and half-siblings healthy

→ LOSS TO FOLLOW UP from age 13-23 years

Age 23

- Sought follow-up with paediatric cardiologist
- Dyspnoea on exertion
- Daily palpitations, worse with exertion

- Social history
- Smoker 15/day, binge EtOH on weekends
- Engaged, not planning a pregnancy, taking oral contraception
- Working sedentary job at supermarket

Issues age 23

1. Failing systemic ventricle
2. Moderate left AV valve incompetence (Ebsteinoid), small VSD
3. Arrhythmia

Discussion re future surgical options:

- TV repair
- PA banding, with view to double switch
- Heart transplantation

12 months later

- Overall symptomatic improvement – no exertional dyspnoea on flat, palpitations reduced in frequency
- Reduced smoking and stopped EtOh intake
- Stopped working, enrolled in college

TTE:

- Mod + TR
- Ventricular EDD 76 mm in short axis
- Mild sub-PS, small perimembranous VSD

LOSS TO FOLLOW-UP Age 24-27

ACHD Follow-up

- Age 27 years
- Presented to district hospital with decompensated heart failure
- Referred to ACHD cardiologist
- Stopped medication for 12 months
- Orthopnoea, PND, dyspnoea at rest
- Palpitations – VT on Holter
- Symptomatic benefit after commencing Carvedilol, Frusemide, Perindopril

- Working as a nursing aid
- Recommenced smoking – ½ packet/day
- Considering pregnancy – off contraception for 12 months

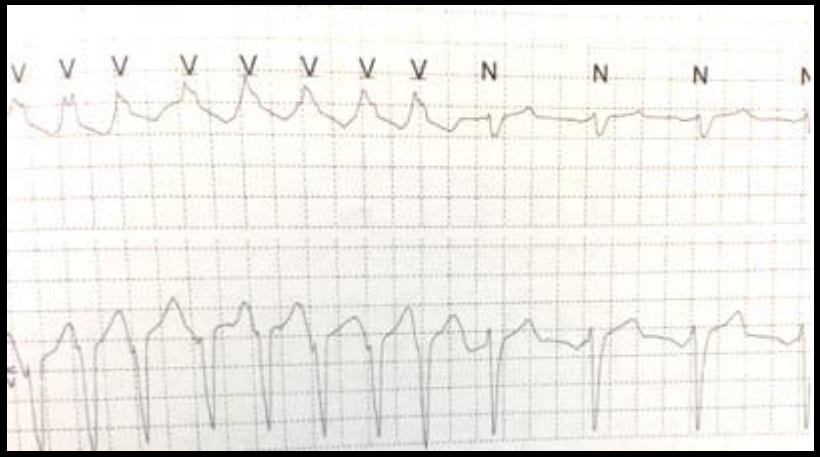
→ Cardiac Catheterisation for PVR assessment

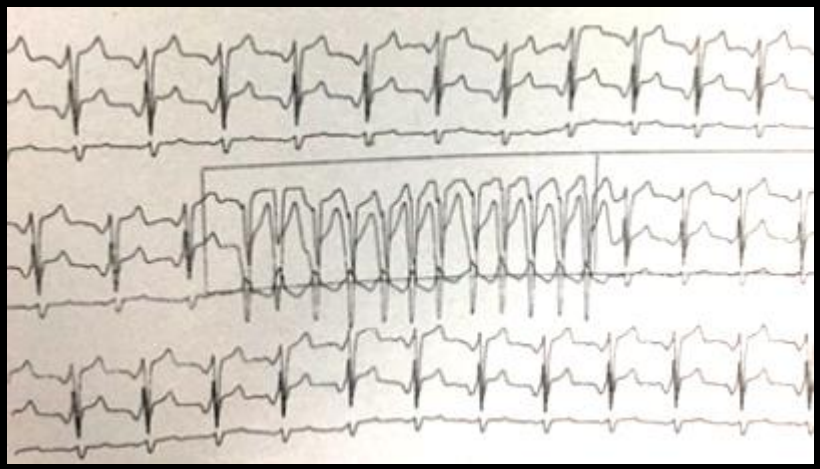
Cardiac Catheterisation 2008

Pressures	(mmHg)	Saturation	(%)	Calculation
RA	7/6/4	SVC	65%	Qp:Qs = 1
Sub-pulm LV	40/4	IVC	65%	
PA	40/12 mean 30	Mid RA	68%	
PCWP	-	MPA	67%	
Systemic RV	120/5-16	Ao	98%	
Aorta	120/60 mean 80			

Echo:
Dilated systemic RV with moderate diffuse hypokinesis
Severe systemic AV valve regurgitation
Giant LA
Mildly elevated PA pressure
Small VSD seen

Arrhythmia





- EP study 2009
- Inducible atrial flutter
- No inducible VT
- Atrial flutter ablation (CTI), atypical flutter still inducible, not ablated.

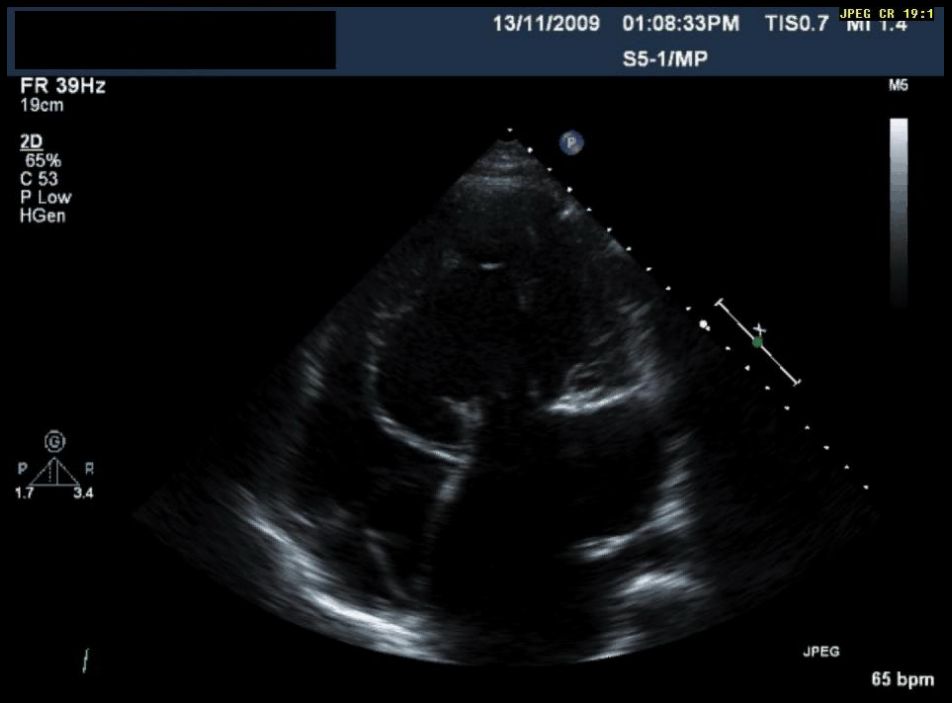
- Dual chamber AICD Insertion
- Endocardial A lead, epicardial V leads with pericardial patch, subxiphoid incision

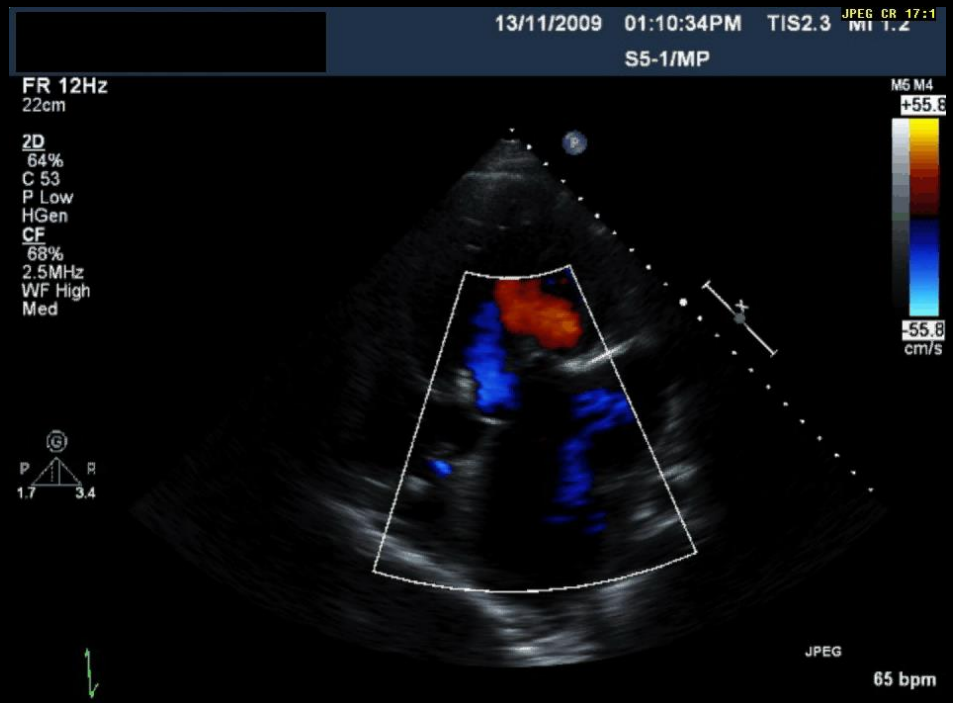
→ “Pericarditis” – oxycontin

Follow-up

- Progressive exertional dyspnoea
- Severe systemic AV valve regurgitation
- Weight gain, BMI 30kg/m^2
- Strongly considering pregnancy
- Continues to smoke
- No contraception

Echo


13/11/2009 01:08:33PM TIS0.7
JPEG CR 19:1
S5-1/MP
FR 39Hz
19cm
2D
65%
C 53
P Low
HGen
M6
P R
1.7 3.4
JPEG
65 bpm


13/11/2009 01:10:34PM TIS2.3
JPEG CR 17:1
S5-1/MP
FR 12Hz
22cm
2D
64%
C 53
P Low
HGen
CF
68%
2.5MHz
WF High
Med
M5 M4
+55.8
-55.8
cm/s
P R
1.7 3.4
JPEG
65 bpm

Valve Replacement Options

- Ebsteinoid tricuspid valve – unreliable repair
- Bioprosthetic valve – if patient considering pregnancy, need for second procedure +/- transplantation as next procedure
- Mechanical valve – concern re thromboembolism – large LA and hypokinetic systemic RV

  - pregnancy and anticoagulation

- Discussed transplantation – too well to consider referral, patient quite opposed to idea

Elective systemic AVVR -30 yo

- Pre-operative Levo
- Dense adhesions
- Ebsteinoid tricuspid valve
- Difficult access → valve replacement 31mm ATS valve

- Uncomplicated post-operative course
- Stable INRs post discharge
- Compliant with warfarin
- Persistent chest wall pain → Oxycontin/Oxynorm

Age 30-33

- Stable
- Attended follow-up, compliant with weight loss (65kg), exercise, smoking cessation, warfarin
- Still strongly seeks pregnancy – no contraception for years
- Perindopril stopped in preparation
- AICD checks – no VT/ICD discharges.

Age 33

- Close relative passes away due to leukaemia
- Stopped exercising, weight gain 78kg
- Smoking, withdrawal method of contraception
- INR control subtherapeutic
- Drug and alcohol problems - metamphetamine, opiates, benzos

Age 34

- Seeks GP assistance for drug addiction
- Admitted to rehabilitation unit for one week
- Discharged to outpatient D&A services after one week

Age 35

- Coronary (trop 360) and cerebral embolism
- Transient speech deficit – complete resolution
- Non-compliant with INR checks – INR =1
- CT – embolic stroke
- TOE – mobile echodensities on the atrial aspect of the mechanical AV valve – 6x2mm, 2x1mm.

→ Anticoagulation continued

Age 35-37

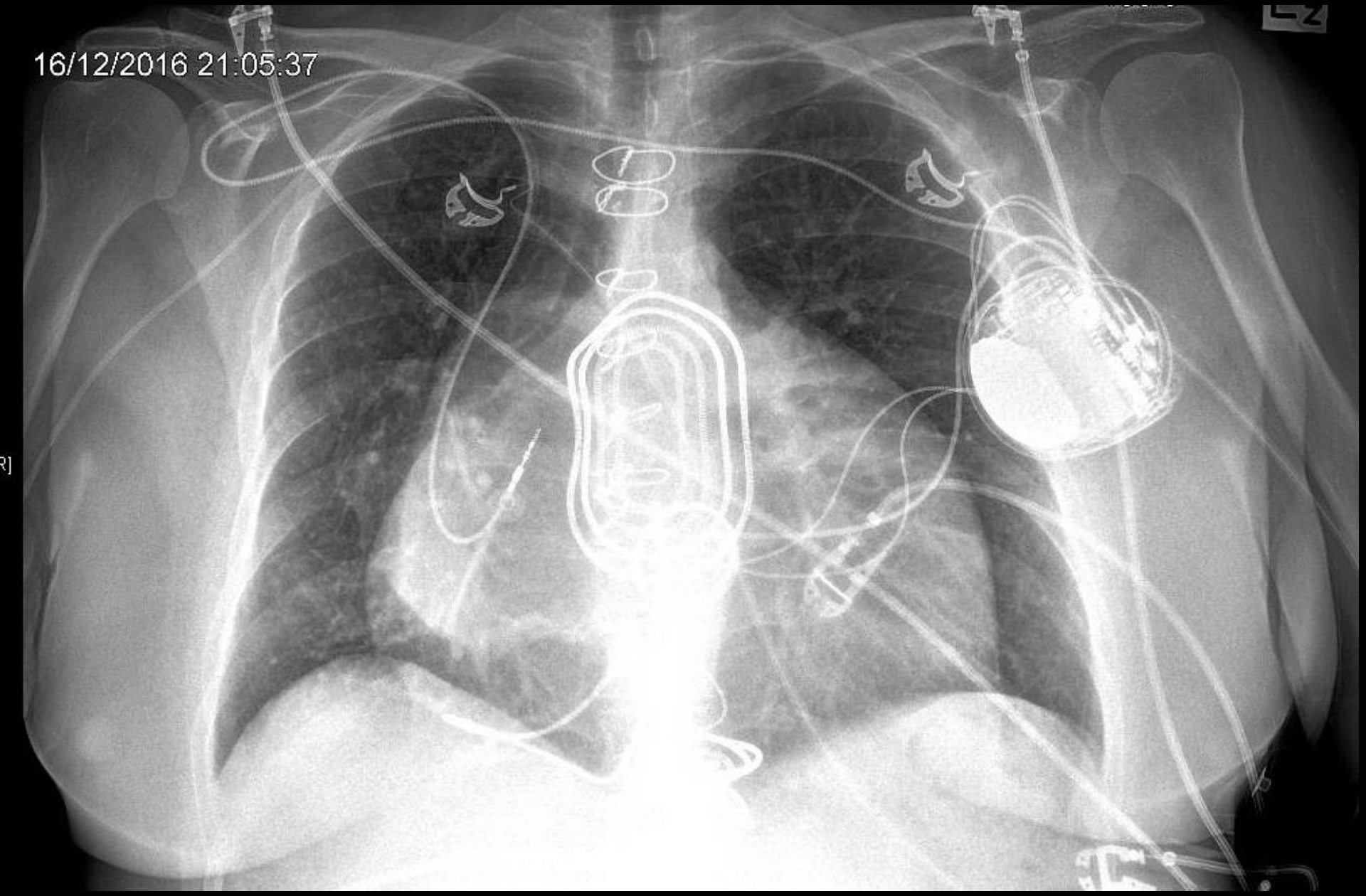
- Loss to follow-up again
- No device checks
- Continues meds (sort of)
  - Frusemide
  - Carvedilol 25mg BD
  - Perindopril 2.5mg
  - Warfarin

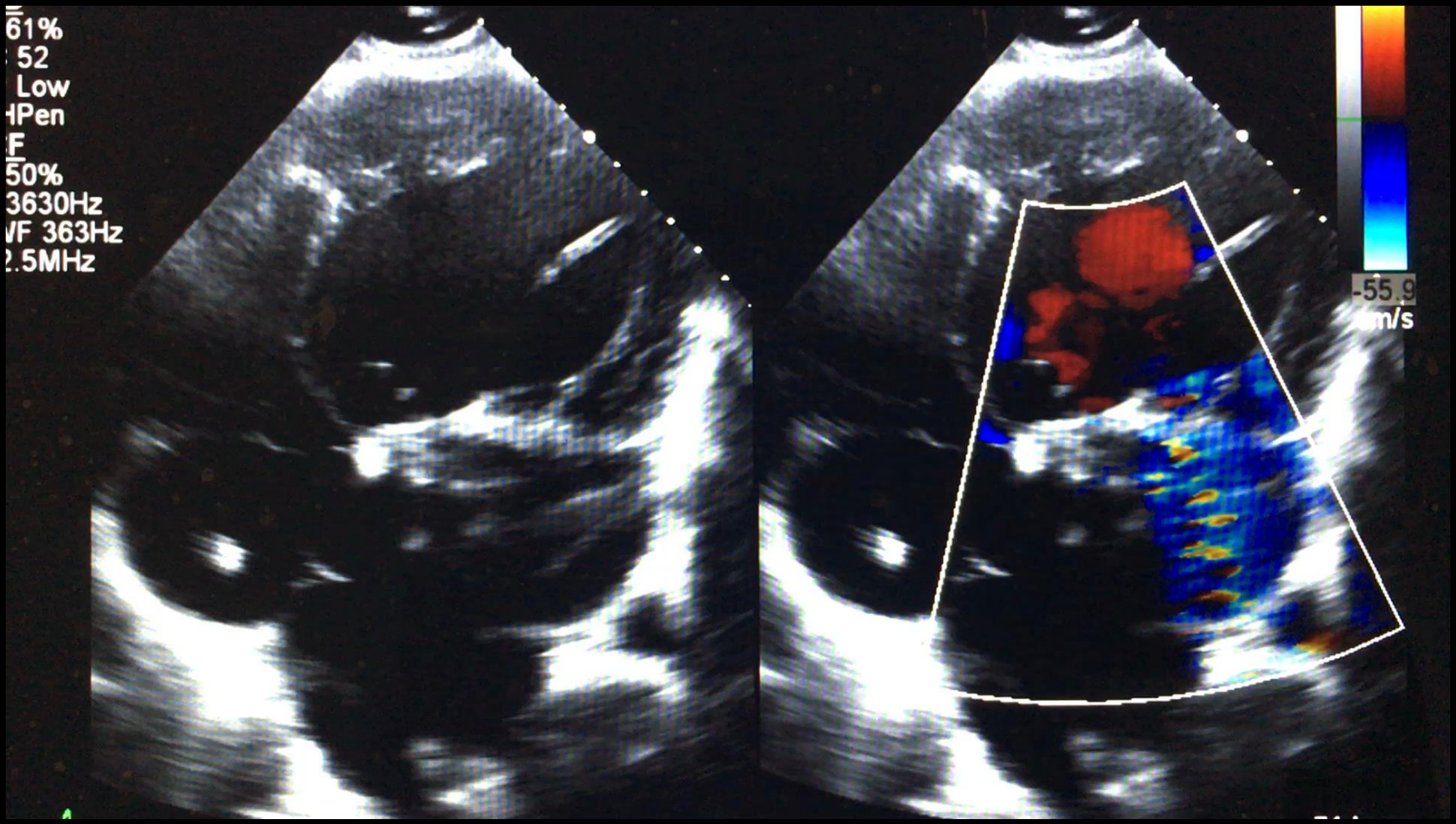
- Exercise intolerance - 1 flight of stairs, NYHA II
- Occasional orthopnoea – self manages with Frusemide 60mg
- Palpitations
- Smokes 1 pack/day

Age 38

- Restarts ACHD follow-up and good GP, declines ψ
  - Venlafaxine
  - Buprenorphine/naloxone
- Severe systemic ventricular impairment, orthopnoea, diuretics uptitrated

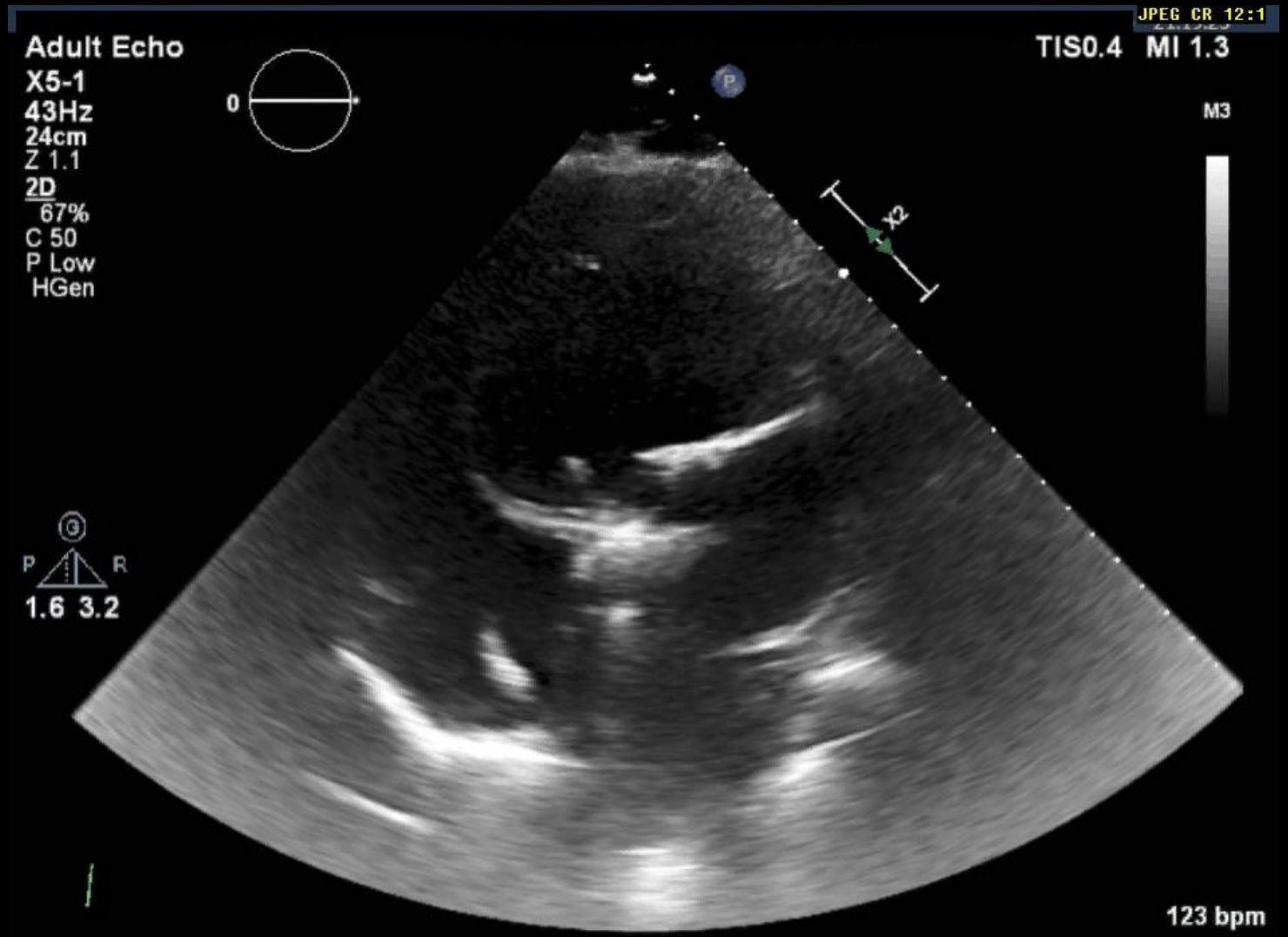
- AICD box change
- VO2 peak = 11 ml/kg/min
- Lung function = FEV1 1.6l 70% predicted, FVC 2.6l
- Blood group A pos
- Discussed transplantation (with patient and SVH)
- Rediscussed smoking cessation, maintaining compliance, INR checks


16/12/2016 21:05:37


61%
52
Low
HPen
F
50%
3630Hz
VF 363Hz
2.5MHz
-55.9
m/s

Age 39

- Presented to district hospital with AF and HR 140 for 2 days
- IV amiodarone
- Frequent runs of VT
- 1x inappropriate shock


JPEG CR 12:1
Adult Echo
X5-1
43Hz
24cm
Z 1.1
2D
67%
C 50
P Low
HGen
TIS0.4 MI 1.3
M3
1.6 3.2
123 bpm

Age 39

- Cardiogenic shock and ischaemic hepatitis
- Milrinone and Amiodarone infusions

- Cardioversion with ECMO on standby
- Slow VT (150) below detection zone. AICD reprogrammed
- Referred to transplant centre

Age 39

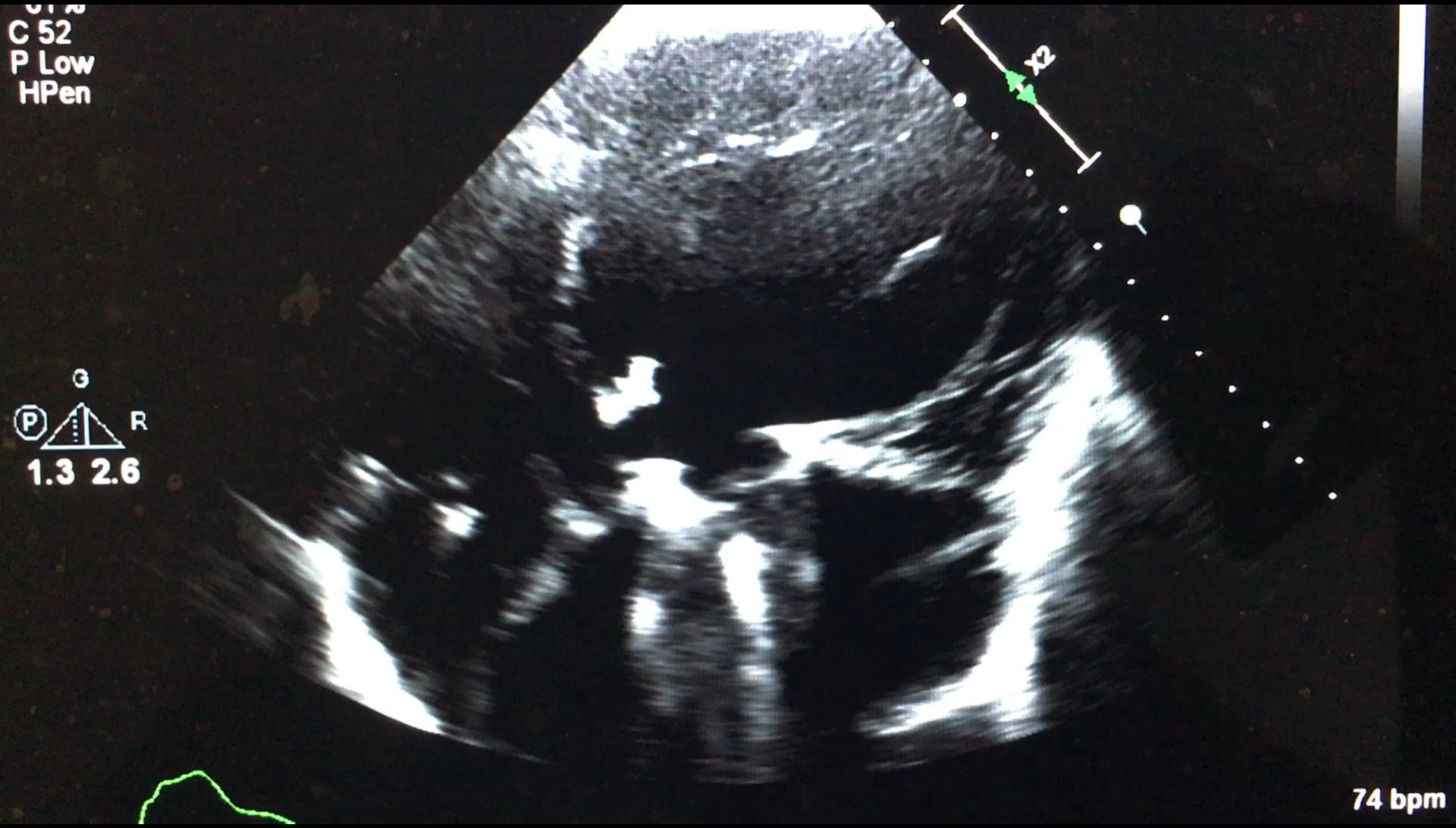
- Stabilised, home
- AICD check – no arrhythmia on amiodarone
- Reviewed at transplant centre
- RHC – VF induced with Swan Ganz in RVOT
- RVSP = 36mmHg – then RHC aborted
- Listed for heart transplant
- Transplanted 1 week later

Progress

- Cardiac transplantation
- Early bleeding
- Developed necrotising pneumonia
- Recurrent air leaks
- ECMO ongoing
- Taken back to address lungs, unable to close chest

Progress

- Treatment withdrawn 2 weeks post-transplant


C 52
P Low
HPen
X2
G
P
R
1.3 2.6
74 bpm