

# Cardiovascular disease in pregnancy

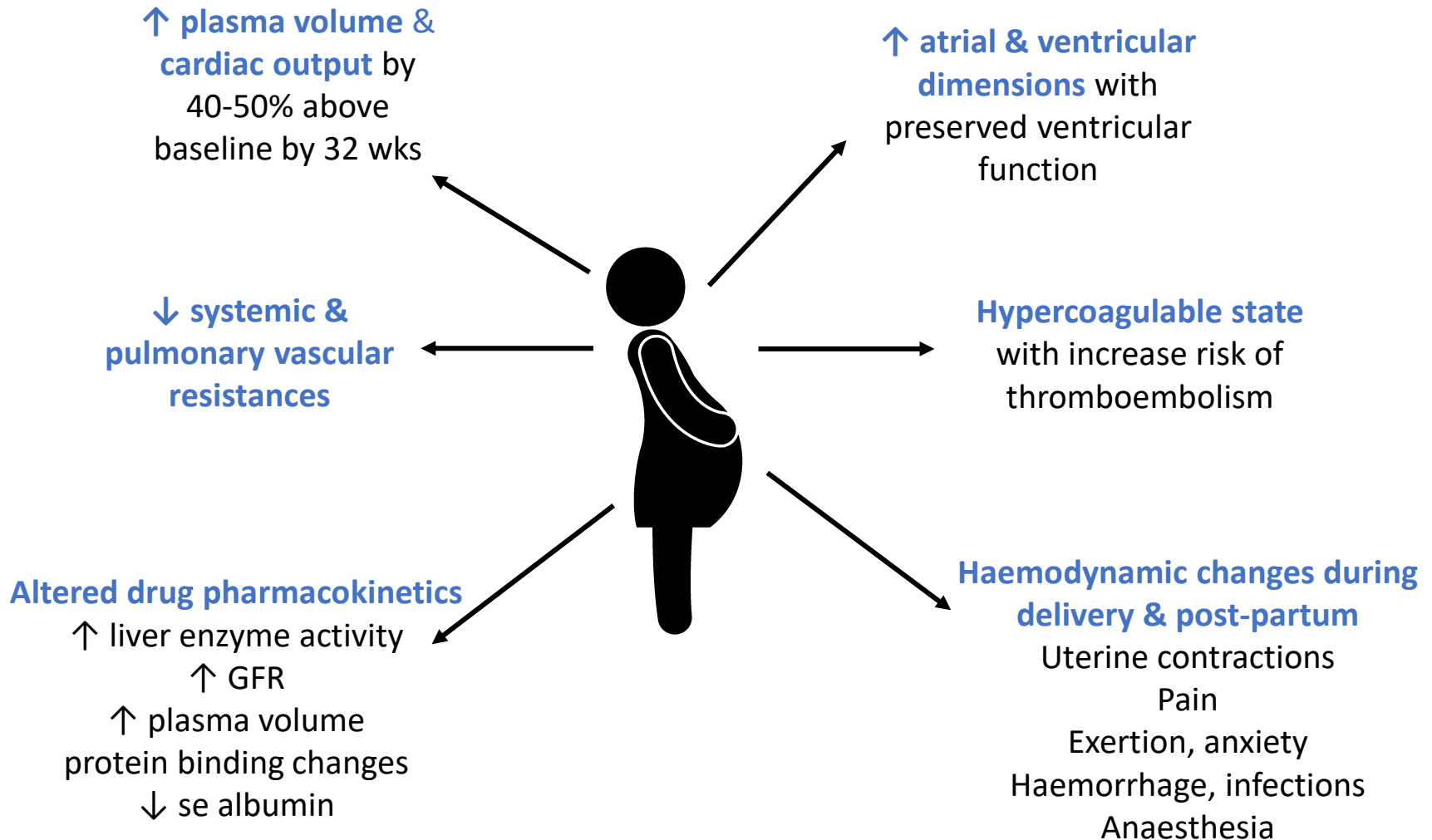
**Dr Maryanne Caruana**

**Consultant Cardiologist, ACHD & Aortopathy Service**

**Dr Tiziana Felice**

**Consultant Cardiologist, Inherited Cardiomyopathy Clinic**

# Physiological adaptations



# Maternal cardiovascular risk stratification

	mWHO class I	mWHO class II
<b>Diagnosis</b>	<ul style="list-style-type: none"> <li>- Small PDA, MVP, mild PS</li> <li>- Repaired ASD, VSD, PDA, anomalous PV drainage</li> <li>- Isolated SVEs/VEs</li> </ul>	<ul style="list-style-type: none"> <li>- Unoperated ASD/VSD</li> <li>- Repaired ToF</li> <li>- Turner syndrome without aortic dilatation</li> <li>- "SVTs"</li> </ul>
<b>Risk</b>	<p>No ↑ mortality No/mildly ↑ morbidity</p>	<p>Small ↑ mortality Moderately ↑ morbidity</p>
<b>Maternal cardiac event rate</b>	2.5 – 5%	5.7 – 10.5%
<b>Minimal f/u during pregnancy</b>	Once / twice	Once per trimester

# Maternal cardiovascular risk stratification

	mWHO class II-III	mWHO class III
<b>Diagnosis</b>	<ul style="list-style-type: none"> <li>- Mild LV impairment (EF &gt;45%)</li> <li>- HCM</li> <li>- Mild MS, moderate AS</li> <li>- Repaired CoA, AVSD</li> <li>- Marfan without aortic dilatation</li> <li>- BAV with aorta &lt;45mm</li> </ul>	<ul style="list-style-type: none"> <li>- Moderate LV impairment (EF 30-45%)</li> <li>- P/H of PPCM without residual LV impairment</li> <li>- Mechanical valve</li> <li>- Uncomplicated + asymptomatic Fontan</li> <li>- Moderate MS</li> <li>- Asymptomatic severe AS</li> <li>- Aorta 40-45mm in Marfan; 45-50mm in BAV)</li> <li>- VTs</li> </ul>
<b>Risk</b>	Intermediate ↑ mortality Moderate/severe ↑ morbidity	Significantly ↑ mortality Severely ↑ morbidity
<b>Maternal cardiac event rate</b>	10 – 19%	19 – 27%
<b>Minimal f/u during pregnancy</b>	Bimonthly	Monthly / bimonthly

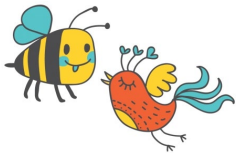
# Maternal cardiovascular risk stratification

	mWHO class IV
<b>Diagnosis</b>	<ul style="list-style-type: none"> <li>- PAH</li> <li>- Severe LV dysfunction (EF &lt;30% / NYHA class III/IV)</li> <li>- P/H of PPCM with residual LV impairment</li> <li>- Severe MS</li> <li>- Severe symptomatic AS / asymptomatic with impaired LV</li> <li>- Marfan aorta &gt;45mm; bicuspid aorta &gt;50mm)</li> <li>- History of aortic dissection</li> <li>- Severe (re)coarctation</li> <li>- Fontan with complications</li> <li>- Vascular Ehlers-Danlos</li> </ul>
<b>Risk</b>	<p>Extremely ↑ mortality Severely ↑ morbidity</p>
<b>Maternal cardiac event rate</b>	40 – 100%
<b>Minimal f/u during pregnancy</b>	Monthly

Pregnancy **CONTRAINDICATED!**  
 Fertility treatment **CONTRAINDICATED!**  
 (Discuss termination if pregnancy occurs)

# Pre-pregnancy counselling

- Informed maternal decision making
- Need for careful pregnancy planning esp. in high risk patients
- Pre-pregnancy tests:
  - ECG, TTE, Exercise test
  - +/- CT/MRI for complete aortic imaging in aortopathies
- Discussion of:
  - Long-term prognosis
  - Fertility & miscarriage rates
  - Risk of recurrence of cardiac disease in offspring
  - Estimates of risk to mother and foetus
  - Medication use in pregnancy
  - Follow-up during pregnancy; delivery plans
  - Encouraging smoking cessation, low alcohol consumption, weight management



# Contraception



- Advice from time of menarche!
- Method of choice = reliability & potential for complications
- *Barrier methods* – **unreliable if used alone**
- *Hormonal*:
  - Ethinyloestradiol-containing contraceptives - high thrombotic risk and can increase BP
  - Progestin-only contraceptives safer in circulations at higher thrombotic risk
    - Oral desogestrel
    - Subdermal implants
    - Levonorgestrel-releasing intra-uterine device (Mirena®)
    - *Sterilisation* - in some specific scenarios

# Medication use in pregnancy

Drug/class	Use in pregnancy	
ACE inhibitors	X	Breastfeeding possible
ARBs	X	Breastfeeding possible
Aspirin (low-dose)	✓	
Adenosine	✓	
Amiodarone	X	
Atenolol	X	
Bumetanide/ frusemide	✓	Oligohydramnios, growth retardation
Carvedilol / metoprolol / propranolol	✓	Hypoglycaemia & bradycardia in foetus



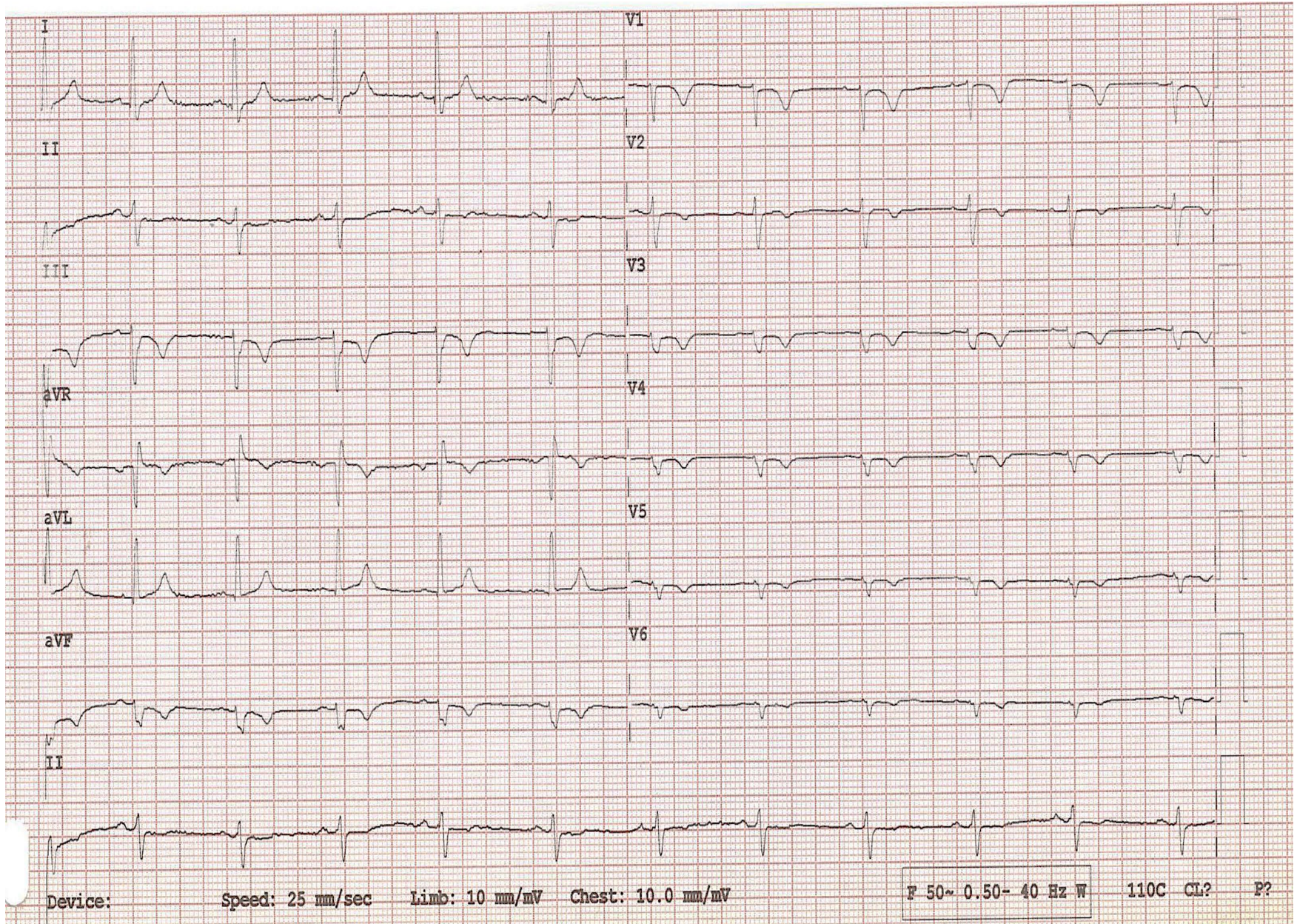
# Medication use in pregnancy

Drug/class	Use in pregnancy	
DOACs	X	Inadequate data
Digoxin	✓	
Flecainide	✓	Prevention of 'SVT' in WPW
Labetalol	✓	
Methyldopa	✓	
Propafenone	✓	Prevention of 'SVT' in WPW
Verapamil	✓	
Warfarin	X/✓	Embryopathy in 1 <sup>st</sup> trimester (dose-dependent)

# CASES

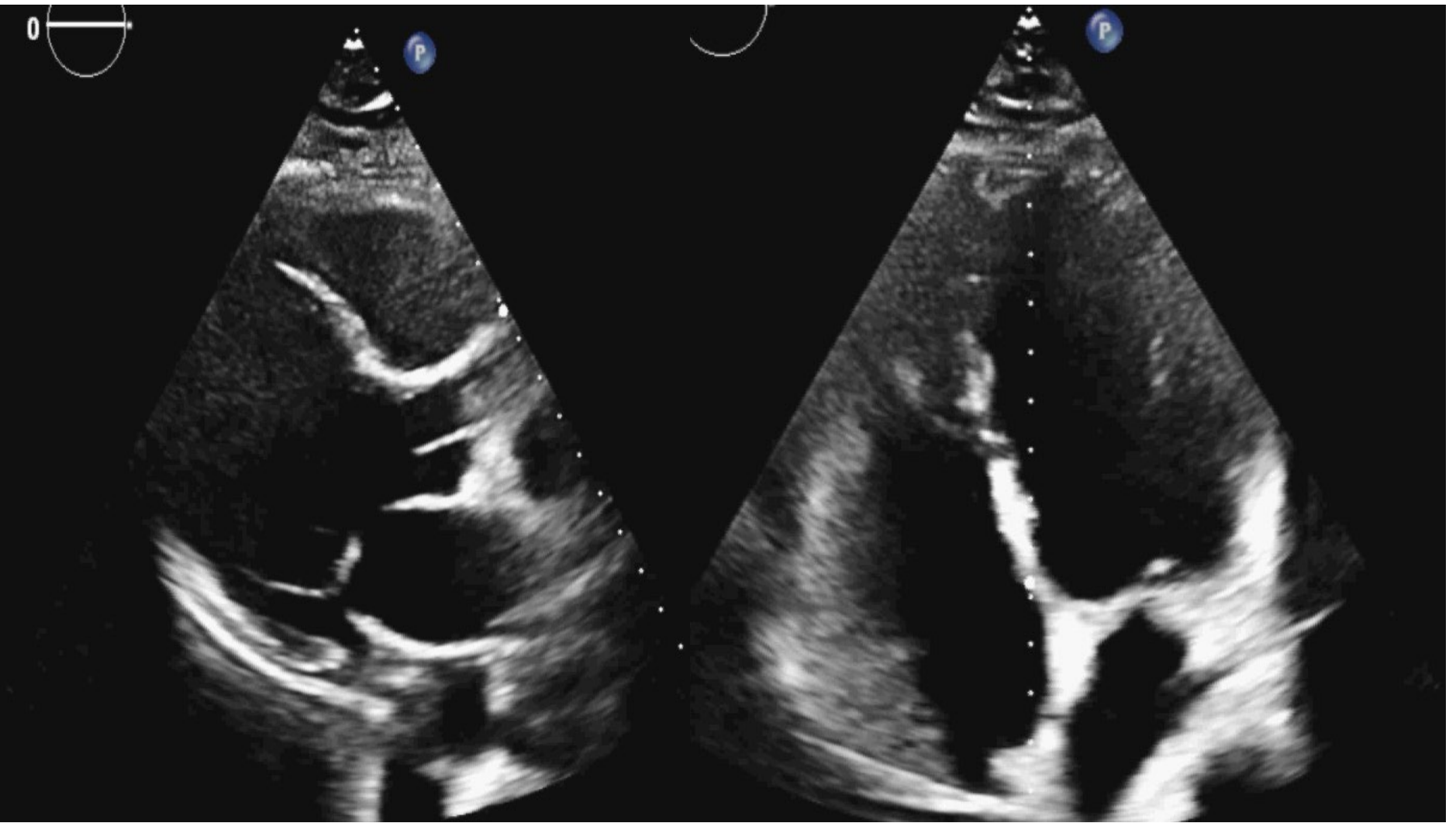
# Case 1

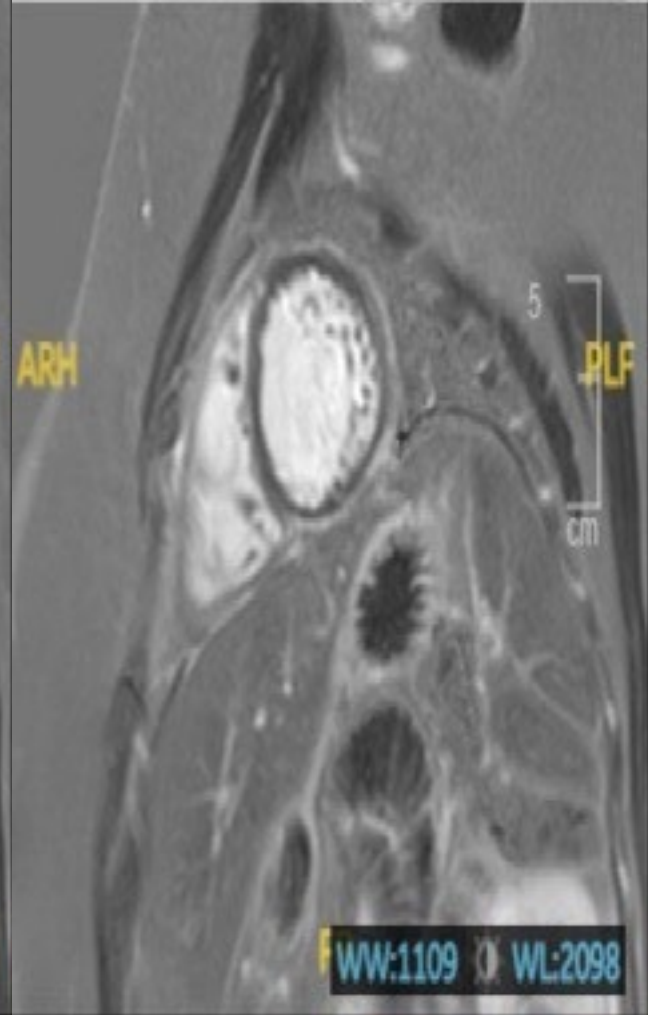
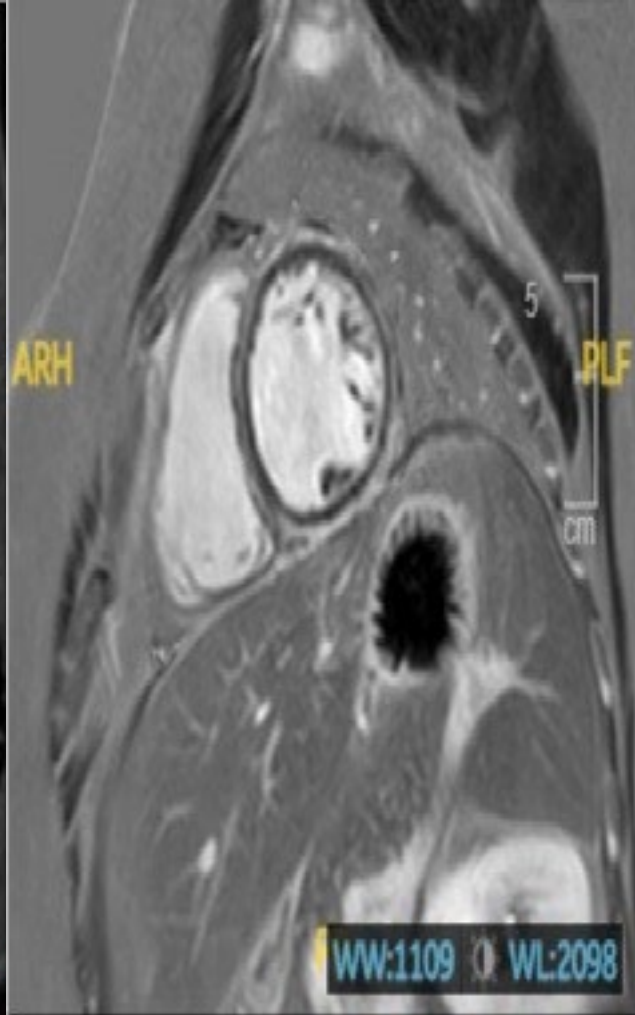
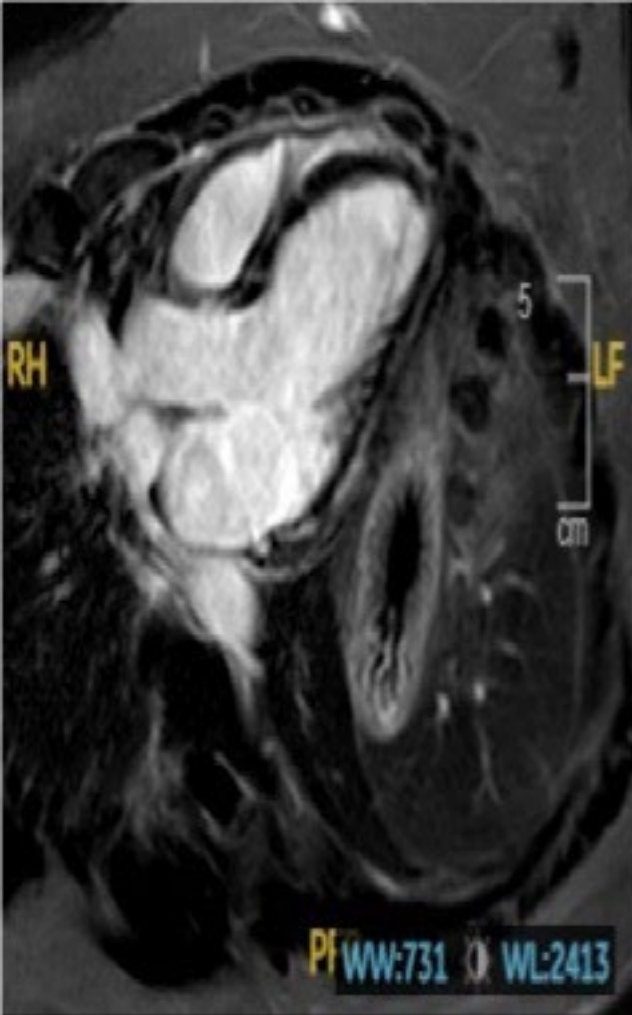
- 28-year-old female referred for cardiac screening: father died suddenly at 40 years of age.
- Asymptomatic
- Examination: Unremarkable
- BMI: 39kg/m<sup>2</sup>



# How would you interpret this ECG?

- A. Normal in view of increased BMI
- B. Suggestive of IHD
- C. Abnormal needs further investigation
- D. Abnormal but asymptomatic, so no further investigations





- EF of 40% and normal LV wall thickness of 8mm
- Circumferential mid-wall and subepicardial late gadolinium enhancement (LGE) in the basal to mid-ventricular LV free wall segments, consistent with a non-ischaemic aetiology

# Diagnosis

- **Genetic testing:** Heterozygous for pathogenic Filamin C variant (c.7384+1G>A)
- **Diagnosis:** Filamin C non-dilated left ventricular cardiomyopathy (NDLVC)



# Management

- Optimised on heart failure medication: Carvedilol, enalapril, spironolactone and empagliflozin, Oral contraceptive pill
- Ejection Fraction improved on treatment
- Genetic and clinical screening of first-degree relatives done
- Paternal uncle genotype and phenotype positive

# Risk Assessment

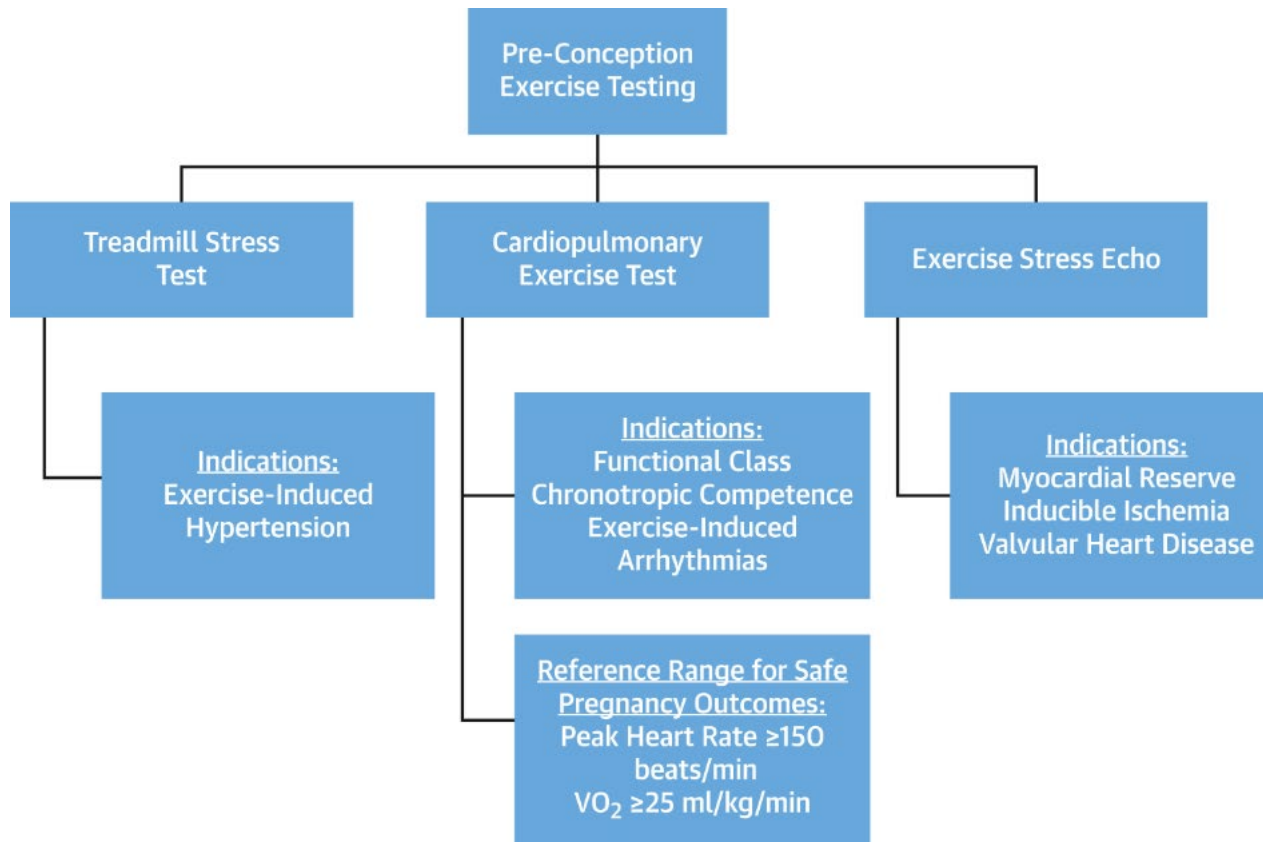
- 24Hr holter: Increased ventricular ectopy.
- Ejection Fraction improved to 45-50%
  
- Risk for SCD:
  - Malignant genetic variant
  - Significant scar

Implantable cardiac defibrillator inserted

# Problem

Patient wishes to become pregnant .....

# Pre-Pregnancy Assessment



# WHO Pregnancy Risk Classification

---

**mWHO I**      Small or mild: pulmonary stenosis, PDA, mitral valve prolapse  
Successfully repaired simple lesions (ASV, VSD, PDA)  
Atrial or ventricular isolated ectopic beats

**mWHO II**      Unoperated ASD/ VSD  
Repaired tetralogy of Fallot  
Most arrhythmias  
Turner syndrome without aortic dilatation

**mWHO II-III**      Mild left ventricular impairment (EF >45%)  
Hypertrophic cardiomyopathy  
Mild mitral stenosis, moderate aortic stenosis  
Marfan syndrome without aortic dilatation  
Aorta <45 mm in bicuspid aortic valve pathology  
Repaired coarctation  
AVSD

# Pre-Pregnancy Assessment

- mWHO Class II-III
- 10-19% maternal cardiac event risk
- Physical limitation on cardiopulmonary exercise test
- Extensive LGE enhancement on MRI

# What's your advice?

- A. Pregnancy is definitely contraindicated
- B. Stop all heart failure medication and allow to become pregnant
- C. The patient has an ICD, pregnancy is contraindicated
- D. Explain the risks of pregnancy. Start removing the HF medications contraindicated in pregnancy

# Pre-Pregnancy Management

- Risk assessment and stratification
- Stop HF drugs CI in pregnancy slowly and repeat echocardiogram in 3 months to assess for LV function deterioration
- Modify existing HF medications:
  - ACE-i, ARBs, ARNIs, MRAs & SGLT-2-i CI
  - Beta-blockers safe



# Outcome

- On stopping her HF medication, a repeat echocardiogram showed a deterioration in ejection fraction to 35-40%
- mWHO Class III 19-27% maternal cardiac event risk = significant increase in mortality and morbidity.
- Risks explained. Patient agreed to avoid pregnancy

## Case 2

- 34 year old lady (cousin of previous case)
- Genetically positive: Filamin C genetic variant
- Father affected with the condition
- History of miscarriage in the past
  
- Very anxious wants to get pregnant

# Which investigations would you perform?

- A. Echocardiogram
- B. 24hr holter
- C. Cardiac MRI
- D. All of the above

# Outcome

- Echocardiogram, cardiac MRI and 24hr holter:  
Normal
- No contra-indications to pregnancy

# How will you monitor the patient during pregnancy?

- A. No need for assessment, EF is normal at baseline
- B. Perform an echocardiogram in the last trimester only
- C. Perform an echocardiogram bimonthly
- D. Perform an echocardiogram once every trimester

# Case 3

- 25 year old lady referred as a new case at ICC
- She had a hospital admission with chest pain and a murmur
- Medication: Atenolol

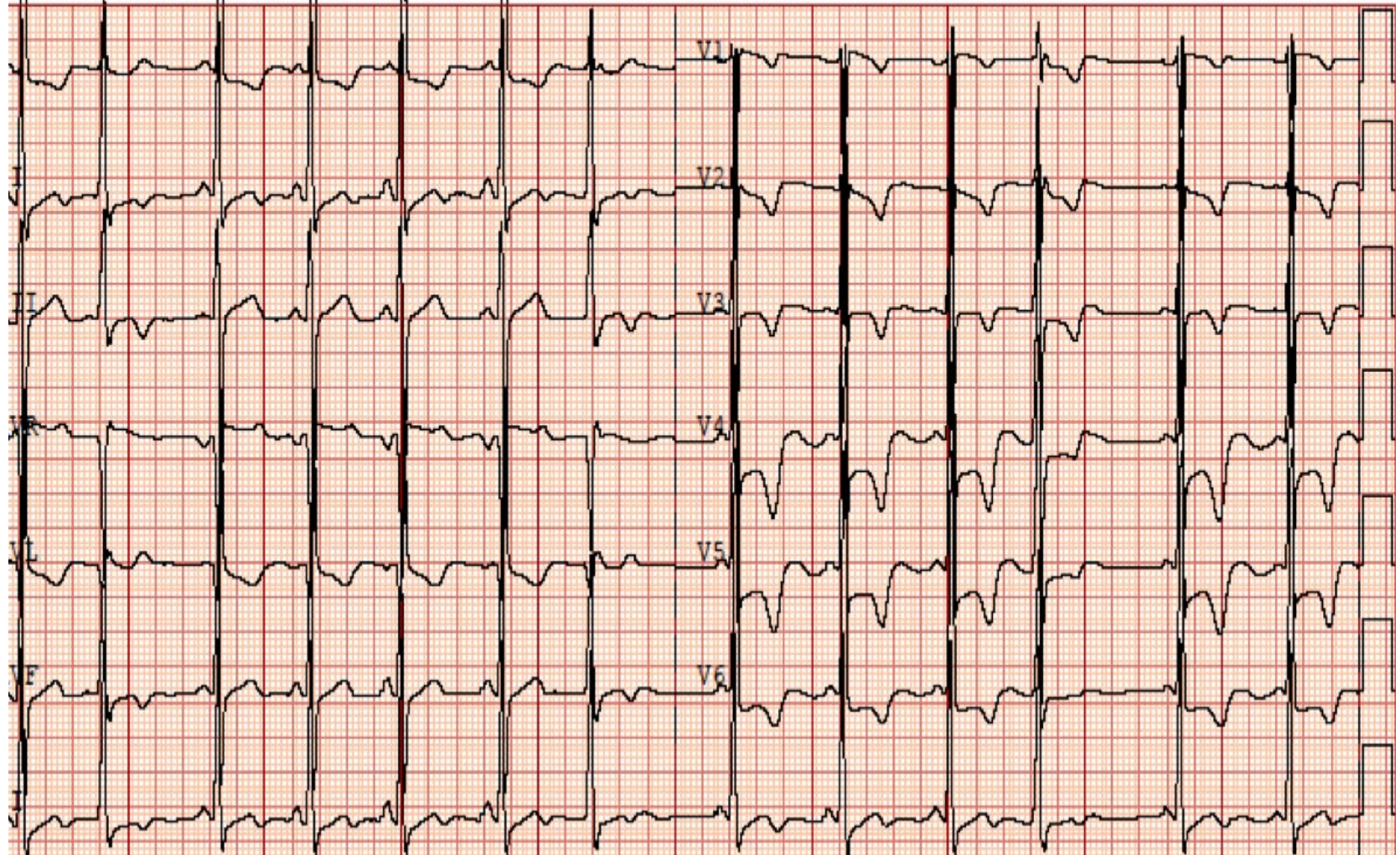
P 48  
QRS 14  
T 143

- ABNORMAL ECG -

Order #: 001YEWQMD  
Enc ID: 001YEWQMD  
Reason: Other

Previous Study: 08-May-2024 04:08:13 - Abnormal Unconfirmed  
L2 Lead; Standard Placement Unconfirmed

Requested By: ^Amy^^Ebejer^

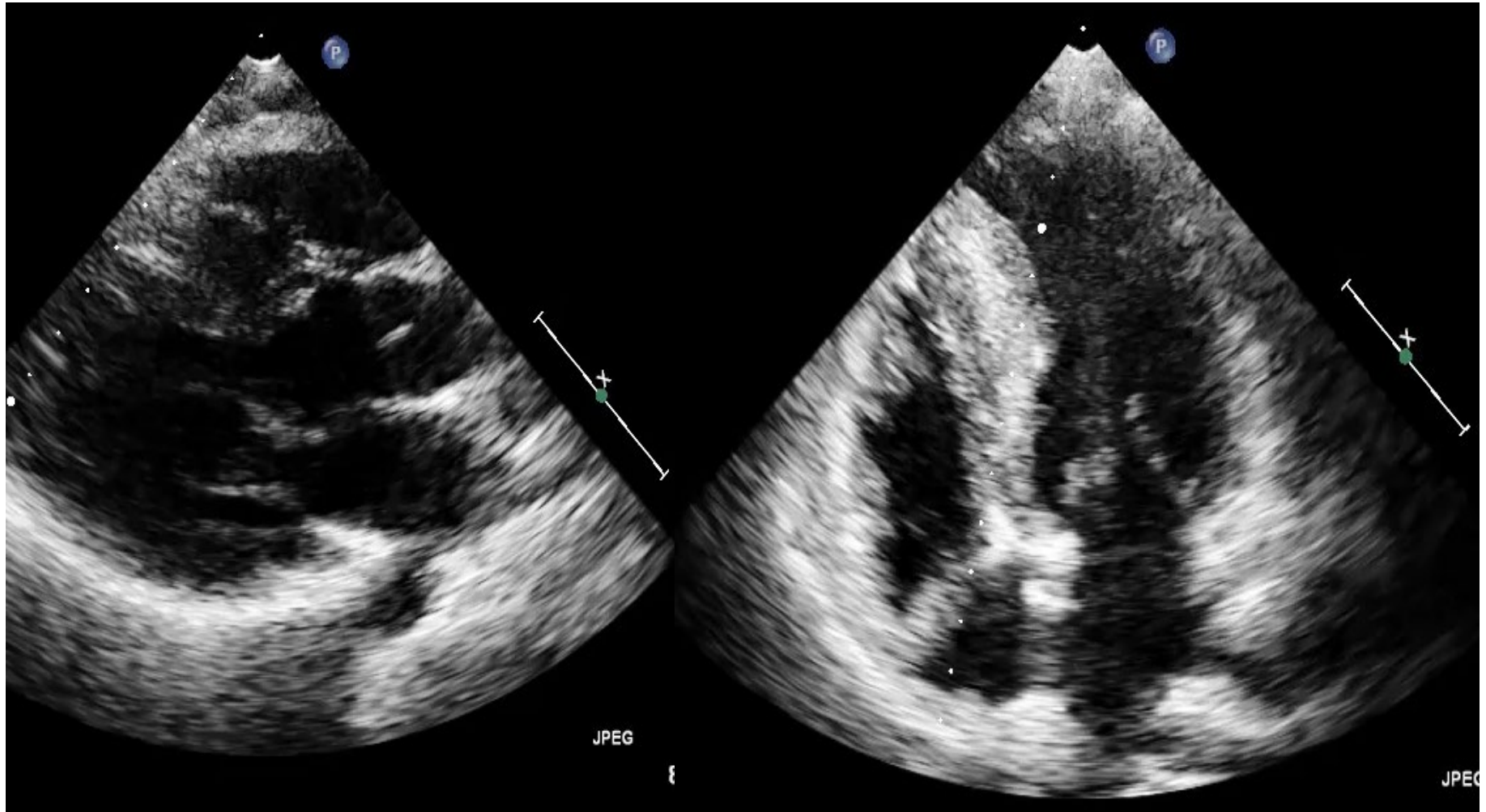


# What does the patient have?

- A. Uncontrolled BP
- B. Acute coronary syndrome
- C. Wolff –Parkinson White
- D. Hypertrophic cardiomyopathy



# Echocardiogram



# Problem

Patient informs me that she is 8 weeks pregnant during first visit

# How will you manage this patient during pregnancy ?

- A. Keep off any medication as she is pregnant
- B. Keep on Atenolol
- C. Change to a more cardio-selective beta-blocker in view of LVOT obstruction
- D. Avoid normal vaginal delivery

# Management and Outcome

- mWHO II-III
- Bimonthly echocardiogram and review
- LVOT gradient remained stable during pregnancy
- Advised to drink plenty of fluids
- Genetically negative
- Anaesthetist and obstetrician informed early on during pregnancy
- Caesarean section successful
  
- Since then she had 3 children in total, with no complications

# Conclusion

- Pre-pregnancy patient-centred counselling and risk estimation are the most important steps
- Management involves further investigations and pharmacological changes accordingly
- MDT approach from pre-conception to postnatal period