

Can we save a woman in a rusted boat with holes,  
loaded with luggage with torn sails and no oars,  
left in the sea, heading towards a storm?

# Case details

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# Mrs. P, 50 years, G2P1L0

House wife  
W/O Mr.M, 64yrs  
From Kurnool  
Class IV SES

- IVF-ICSI conception with Donor embryo (D5)
- She was referred on 4<sup>th</sup> July by her primary consultant
- She came 2 days later to FH at 29 weeks, with
  - H/o SOB ↑ in the last 1-2 weeks
  - H/O Orthopnoea
  - No H/O PND or reduced urine output

LMP:21.12.18

EDD:28.9.19

Corrected EDD (D5 ET) : 21.9.19

Conscious, oriented, mild pallor+ pedal edema G II JVP not elevated

PR: 102/mt RR: 24/mt BP: 160/90mHg SPO2: 95% (5l O2)

CVS: S1S2 + Systolic murmur +; RS : B/L Basal crepitations +

Booking BMI  
36.34kg/m<sup>2</sup>  
? weight gain **19** kg

- 
- After initial assessment she was transferred to Virinchi Hospital for joint expert care
  - Baseline cardiac evaluation and work up was done
  - Hospitalization & Decongestive therapy → improved to NYHA II and was discharged after 4 days
  - Was advised to continue the decongestive therapy in addition to labetalol

# Investigations during first visit

CBP – Hb 10.5gm / 7500 / 2.5 lakh per cu mm

CUE – NAD

LFT – normal except Albumin 2mg/dl

S. Creatinine 0.5mg/dl

S. Electrolytes normal

FBS & PPBS – 89 & 126mg%

TSH – 4.27

PT / aPTT normal

S. Calcium 7.2mg%

6/7/19

RHD, Severe MS/MR,  
Mild TR, Modertae AR RVSP 43

L atrial dilatation+

EF 55%

No effusion

No clot/vegetation

ECG

1. Tab. Furosemide 40 mg twice daily
2. Tab .Labetalol 100mg Q 12<sup>th</sup> hourly
3. Tab. Levothyroxine 25 mg
4. Sry.Potchlor and Mucaine gel
5. Iron, Calcium and Vit D

# Second visit to us at 30<sup>+3</sup> weeks for Physician review

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- SOB on day to day activities -7 to 10 days
- H/O orthopnoea - 7 to 10 days
- ↑Epigastric pain - 10 days
  
- No H/O PND, reduced urine output, chest pain, palpitations
  
- No imminent symptoms
  
- Compliance → poor → taking labetalol 100mg OD

# Obstetric History

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- First pregnancy – seven years after marriage, Spontaneous conception  
Uneventful SVD of ? AGA baby, Neonatal death D 15 ? Cause
- Second pregnancy – present one ; IVF donor embryo
- No details about earlier visits and investigation reports

## Medical History:

- Hypertension since 2018
- Hypothyroid during IVF 2018

## Surgical history :

- Underwent laparoscopy in 1995
- Hysteroscopy in 2018 before IVF pregnancy

Family history : No family H/O HTN, DM, IHD, sudden death, twins, malignancies

## Treatment History :

- Was on Amlodipine 5mg OD changed to labetalol 100mg TID
- Levothyroxine 25mcg OD

# Examination

- Comfortable, not tachypnoeic / dyspnoeic
- Mild pallor+, Pedal edema Grade II, no icterus, no cyanosis, No lymphadenopathy & temperature normal
- JVP not elevated
- B/L legs → edema, thick & dry skin with scratch marks
- PR: 84/ mt, regular, normal volume, all peripheral pulses felt
- BP: 160/100mmHg → 140/92mmHg in Left arm, sitting position
- Uterus 34 weeks, relaxed, multiple fetal parts felt, liquor adequate, 1<sup>st</sup> cephalic, Both FHR localized
- No hepatomegaly

155 cm

Breast, Thyroid, Spine Normal



# CVS

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- Woman in lying position with 30-45° incline
- Apical impulse on fourth intercostal space little lateral to mid clavicular line
- No visible pulsations

On palpation, apical impulse → heaving

- No parasternal heave
- No thrill

On Auscultation,

- S1 S2 heard
- Pansystolic murmur of Grade IV
- Short systolic murmur in pulmonary area
- Systolic murmur in para sternal area
- No murmur in aortic area
- No S3 or gallop rhythm

# Diagnosis

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G2P1L0 30<sup>+3</sup> weeks twin gestation, Chronic HTN, GDM (D), Hypothyroid, Anemia (mild), RHD MS(severe) MR/TR/AR mild pulmonary hypertension NYHA class II to III in sinus rhythm not in failure

# RISK FACTORS

- Advanced maternal age ★
- Twin gestation ★
- RHD – MS, MR, PHTN
- Chronic hypertension ★
- Hypothyroidism
- GDM (Diet) – Insulin ★
- Anaemia ★
- ?Fungal infection
- Obesity ★

## CARDIAC RISK ASSESSMENT

### At first admission

Carpreg score - 2  
mWHO class - IV

### Second visit

› Carpreg score - 3  
› mWHO class - III

**Weight gain : 12kg**

# First visit - summary

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- In Acute congestive heart failure
- Maternal stabilization
- Medical measures
- Thorough evaluation
- Functional status improved
- Discharged on medications

# Second visit

**Advised admission till delivery** (Considering her functional status, distance travel and availability of comprehensive expert clinical care)

## Delivery plan was made

- Timing of delivery & Place of delivery
- Steroids
- Neonatal plan

## Maternal evaluation

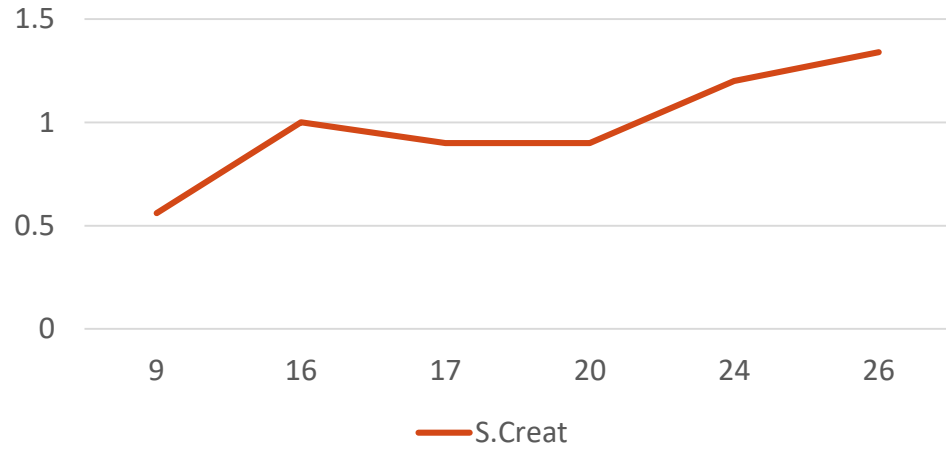
- Work up for Chronic Hypertension –control of BP
- OGTT →Blood sugar monitoring
- Daily assessment of symptoms
- Continued all medications / treat anemia
- High risk consent
- Thromboprophylaxis
- Control of risk factors (many)
- Joint consultation – cardiologist/maternal-fetal medicine/ critical care-anaesthetist/ neonatologist

## Fetal evaluation

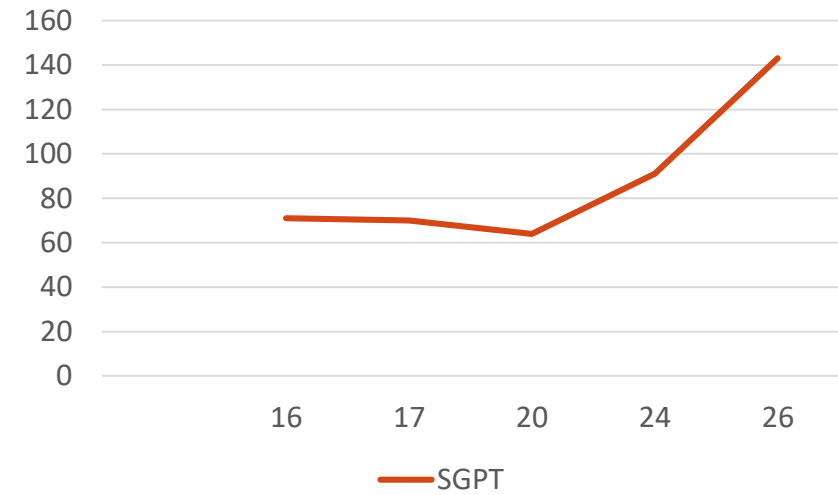
- USG for growth and AFI
- Bi weekly NST
- Daily fetal Kick count
- Corticosteroids
- Neonatal counselling – prematurity, Twins

# Trend in laboratory parameters

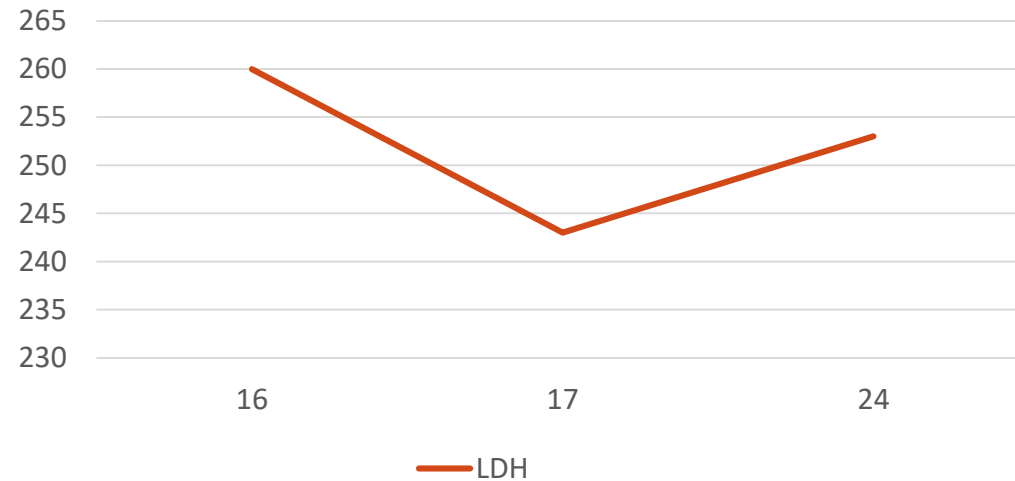
## S.Creat



## SGPT



## LDH



Blood Urea : 63mg%  
S.Bilirubin – 1.06mg%

However after withstanding the storm, the woman requires not only support from her family but also requires extended postpartum care. Need to develop protocol to follow up the woman and support her to go through motherhood fearlessly and access medical care as well

- ❖ There was maternal deterioration (?) in terms of ↑ SGPT, LDH & Creatinine
- ❖ Planned for elective caesarean section at VH at 32 weeks after steroid cover
- ❖ Intra OP – Uneventful; LSCS was done under GA
- ❖ Post OP – intensive monitoring for 2 days
- ❖ Discharged after 5 days
- ❖ Babies in NICU → 1.3Kg (gained 40gms) & 1.2 kg (gained 20gms) doing well as of now-discharged after 10 days

# Evaluation

## Done

- Basic investigations
- S. Electrolytes
- S. Creatinine
- Lipid profile
- ECG
- TT Echo
- USG abdomen
- Renal Doppler
- Fundus examination
- Multidisciplinary care
- Pregnancy plan management

## Time did not permit

- Pre conceptional counselling
- Optimization of cardiac status before ART
- To look for medical risk factors & optimize before pregnancy
- Periconceptional folic acid?
- First trimester screening- prenatal diagnosis / TIFFA
- Early consultation to cardiologist in pregnancy
- Aspirin at the earliest
- Early hospitalization
- Psychological counselling
- Prophylaxis for rheumatic fever



# Discussion

**ADVANCED MATERNAL AGE**

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**MITRAL STENOSIS**

# Advanced maternal age

- Fetal aneuploidy
- Fetal malformation
- DM
- HTN
- APH
- PROM
- Preterm labour
- Cardiac disease

Obesity

Female → 35 years

Male → ? 40 years

- Autism spectrum
- Schizophrenia
- Mutations in FGFR2 &3 genes –skeletal dysplasias & craniosynostosis syndromes

Birth after 35 years → 9 times increased

>40 years → 2.5% of pregnancies (70%)

Multi fetal pregnancies

Increased operative deliveries & still birth rate

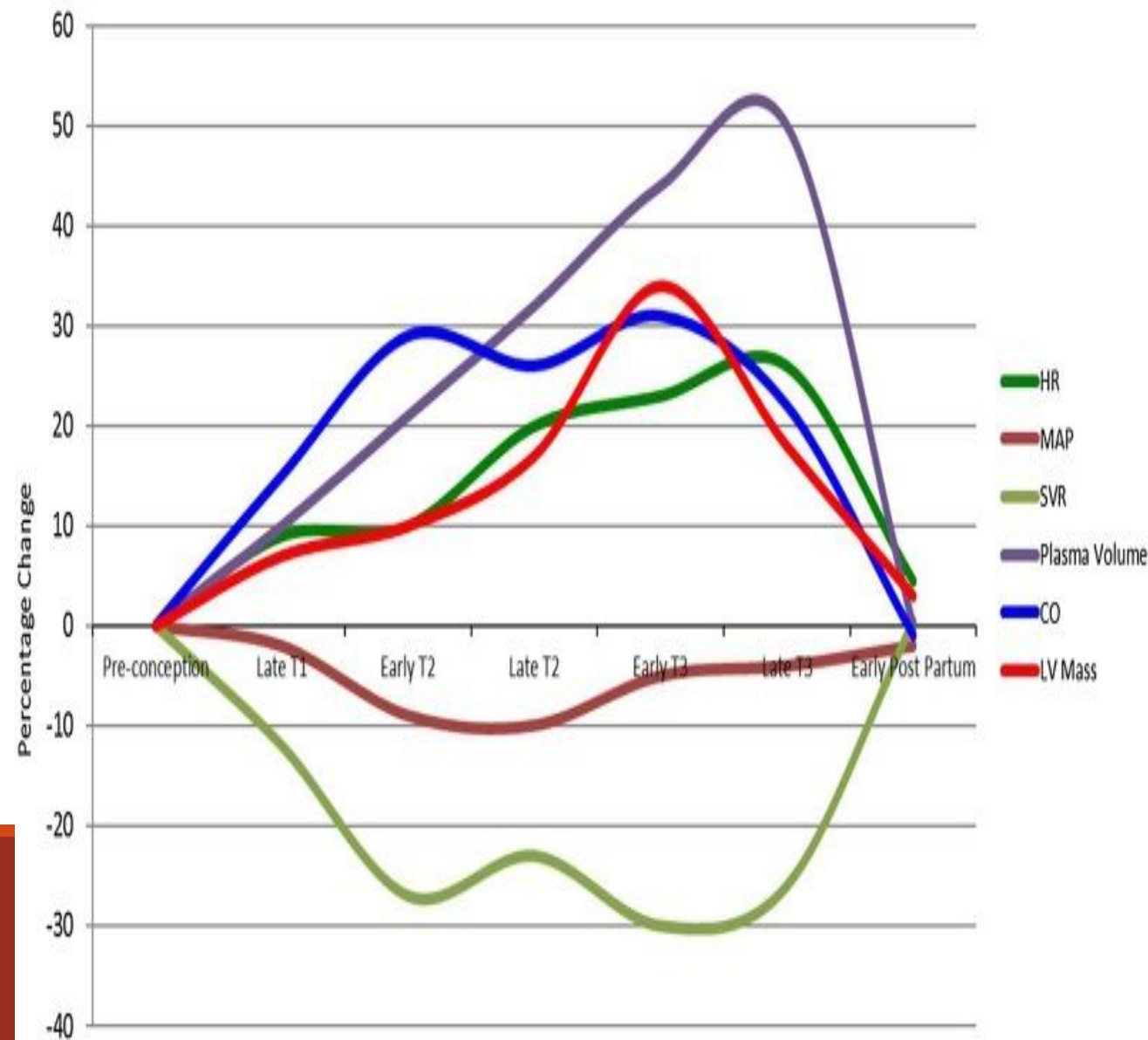
# Cardiac disease in pregnancy

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- Cardiac disease – it affects 1-4% of pregnant women
- Congenital heart disease is on the rise
- Significant morbidity from RHD - developing countries
- Increased number of risk factors – Advanced maternal age, Hypertension, obesity, diabetes mellitus and multiple pregnancies
- Single most important indirect cause of maternal death

- Leading cause of maternal death in developed countries...
- In the UK, the Confidential Enquiries into Maternal Deaths (CEMACH) have shown that the overall rate of mortality from cardiac disease has **tripled** in two decades #
- One-third of these deaths are a result of **MI/ IHD** and a similar number of late deaths are associated with **PPCM**
- A recent study of maternal cardiovascular mortality in Illinois found that 28.1% of maternal cardiac deaths were **potentially deemed preventable** due to health care provider issues, patient features (eg, nonadherence, obesity) and health care system factors related to access\* (**97% acquired HD, 75% cardiac related deaths**)

- CO, HR, Plasma volume & SV
- SVR
- BP
- Structural changes
- The changes are mandatory to cope up with extra circulating blood volume
- Left ventricular mass and vascular resistance do not fully return to pre-pregnancy levels



## Physiological changes

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- Expression of oestrogen receptors in the aorta causes fragmentation of reticulin fibres, reduced amount of acid mucopolysaccharides and loss of the normal arrangement of elastin fibers, predisposing women to aortic dissection, particularly if they have an aortopathy
  - Additionally, pregnancy is a hypercoagulable state, designed to reduce the risk of post-partum haemorrhage → increased risk of TE

# Pre pregnancy counselling & Risk assessment

- ❑ Need to have frank discussions
- ❑ Risks to the mother and foetus
- ❑ Pregnancy and longevity
- ❑ Recurrence rate
- ❑ Assisted reproductive techniques
- ❑ Contraception

- The most important aspect of assessment of reproductive-age women with cardiac disease is pre-conception counselling
- Joint counselling with experts
- Multivitamins and folic acid supplementation
- Modification of cardiac drugs
- Optimization of cardiac status
- Genetic counselling

# Risk assessment

- ❖ History and physical examination, a 12-lead ECG & transthoracic echocardiogram
- ❖ Cardiac CT & MRI if necessary
- ❖ Exercise stress testing to measure functional capacity and BP response to exercise is useful
- ❖ Cardiopulmonary testing & SPO2
- ❖ Baseline and serial serum B-type natriuretic peptide levels during pregnancy can be incorporated into pregnancy assessment in women with the potential to develop heart failure (HF)
- ❖ Women with arrhythmias may benefit from continuous ECG monitoring, exercise testing or electrophysiology studies



# CARPREG I

Siu SC, Sermer M, Colman JM, Alvarez AN, Mercier LA, Morton BC, et al; Cardiac Disease in Pregnancy (CARPREG) Investigators. Prospective multicenter study of pregnancy outcomes in women with heart disease. *Circulation*. 2001;104(5):515-21.

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- Prior cardiac event or arrhythmia
- New York Heart Association (NYHA) Class >II or cyanosis
- Left heart obstruction
- Systemic ventricular dysfunction (ejection fraction <40%)

The multicenter CARPREG (Cardiac Disease in Pregnancy Study) was the first to develop a risk index to predict the likelihood of maternal cardiac complications from general maternal clinical & echocardiographic data obtained during the baseline antepartum visit

0: 5% risk

1: 27% risk

≥ 2: 75% risk

# ZAHARA score

ZAHARA (Zwangerschap bij Aangeboren HARTafwijking

*[Pregnancy in Women With Congenital Heart Disease]*

weighted risk score that included components of the CARPREG risk index

## Score & Risk of Cardiac Complications

0-0.5	2.9% risk
<b>0.51-1.5</b>	<b>7.5% risk</b>
1.51-2.5	17.5% risk
2.51-3.5	43.1% risk
<b>&gt;3.51</b>	<b>70% risk</b>

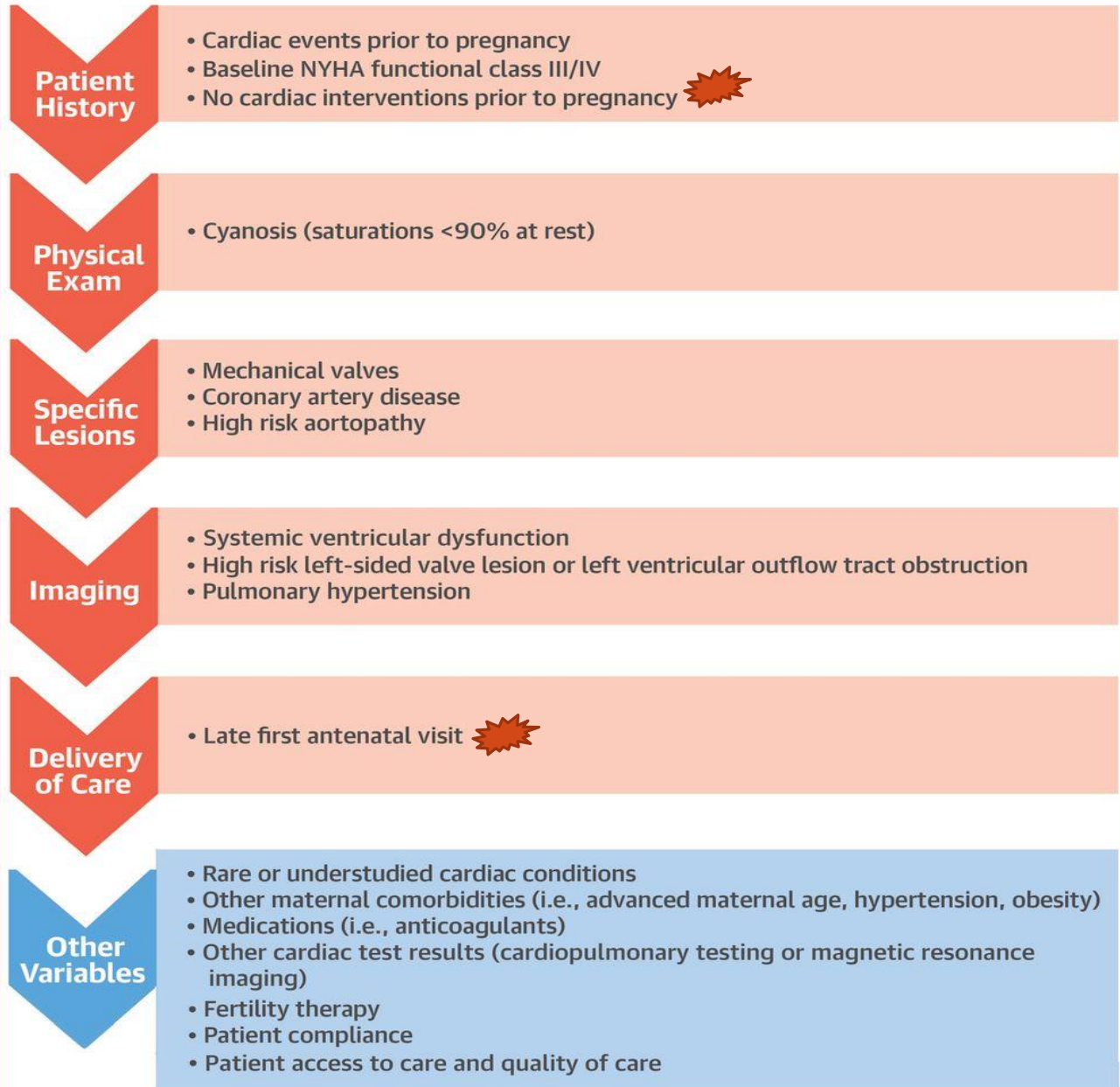
Risk Factor and Weight	Points
History of arrhythmia	1.5
Cardiac medication prior to pregnancy	1.5
NYHA Class $\geq$ II	0.75
Left heart obstruction	2.5
Systemic atrioventricular valve regurgitation (moderate or severe)	0.75
Pulmonic atrioventricular valve regurgitation (moderate or severe)	0.75
Mechanical valve prosthesis	4.25
Cyanotic heart disease (corrected or uncorrected)	1.0

# CARPREG II study

- **5 general predictors** (prior cardiac events or arrhythmias, poor functional class or cyanosis, high-risk valve disease/left ventricular outflow tract obstruction, systemic ventricular dysfunction, no prior cardiac interventions);
- **4 lesion-specific predictors** (mechanical valves, high-risk aortopathies, pulmonary hypertension, coronary artery disease);
- **1 delivery of care predictor** (late pregnancy assessment)

10 predictors of maternal cardiac complications were identified and incorporated into CARPREG II score

## CENTRAL ILLUSTRATION: Predictors of Adverse Events in Pregnant Women With Heart Disease



- In the European Registry of Pregnancy and Cardiac diseases (ROPAC), the prevalence of native MS during pregnancy, either isolated or with mitral regurgitation was 70%; 39.2% of patients had moderate and 19.8% had severe MS
- The prevalence was even higher when including patients with percutaneously or surgically corrected MS
- Delays in recognition of cardiovascular symptoms during pregnancy
- 2015 report from UK → substandard health care accounted for more than 50% of cardiac deaths, half of which were considered avoidable

# mWHO class

- The consensus-based modified World Health Organization (mWHO) classification was proposed to be a more comprehensive risk stratification method
- Agreed worldwide



- The mWHO classification is currently the most accurate system of risk assessment, although it is probably more appropriate for developed, rather than developing, countries
- Indications for intervention (surgical or catheter) do not differ in women who contemplate pregnancy compared with other patients (moderate MS/ aortic dilatation)
- Fertility treatment is contraindicated in women with mWHO class IV, and should be carefully considered in those who have mWHO class III disease or who are anticoagulated

Balci A et al. Prospective validation and assessment of cardiovascular and offspring risk models for pregnant women with congenital heart disease. *Heart* 2014;100:1373-81

## Class I

- No detectable increased risk of maternal mortality
- No/minimal increase in maternal morbidity

## Class II

- Small increased risk of maternal mortality
- Moderate increase in morbidity

## Class II-III

- Intermediate increased risk of maternal mortality
- Moderate to severe increase in morbidity

## Class III

- Significantly ↑ risk of maternal mortality or severe morbidity  
expert cardiac & obstetric pre-pregnancy, antenatal & postnatal care are required

## Class IV

- Extremely increased risk of morbidity and mortality
- Pregnancy is contraindicated

# Mitral stenosis - pathophysiology

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- Most common cause is rheumatic heart disease
- Normal mitral valve orifice area  $4-6\text{cm}^2 \rightarrow 2\text{cm}^2$  classical symptoms of heart disease starts appearing
- Effective penicillin prophylaxis shifted the burden of the disease to third decade
- Most common presentation of RHD is MS followed by MS with AR
- Thickening & immobility of the leaflets, thickening and fusion of the chordae tendinae or mitral annular & commissural calcification

## Symptoms most commonly develop during third trimester and intrapartum

- Echo – diagnostic
  - Auscultation of MDM is diagnostic of MS
  - Critical clinical aspect is heart rate
  - Rate control is essential
- Rapid heart rate shortens diastolic filling time, increases the left atrial pressure and the pulmonary venous pressure and causes heart failure symptoms
  - Patients with MS are dependent on atrial contraction → why development of atrial fibrillation (AF) is problematic
  - About 50% of patients with severe MS will develop heart failure symptoms during pregnancy



# Assessment of severity

Measurement	Normal	Mild	Moderate	severe
Mitral valve area (Cm <sup>2</sup> )	4.0-6.0	1.5-2.5	1-1.5	<1
Mean pressure gradient (mmHg)	<2	<5	5-10	>10
Pulmonary artery mean pressure (mmHg)	10-20	<30	30-50	>50
Pulmonary edema		11-24%	34-61%	56-78%
Atrial fibrillation		0-7%	10-22%	33%

Fetal risks → Prematurity (20-30%), FGR (5-20%) & foetal death (1-5%)

# Interventions & its timings

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Symptomatic moderate stenosis & severe stenosis should ideally undergo treatment before pregnancy

In moderate & severe mitral stenosis, decisions around delivery often need to be made on a week by week basis as pregnancy progresses.

## Medications

- Beta blockers for rate control → lengthen the diastolic filling time & reduce left atrial pressure
- Diuretics as decongestive therapy
- Digoxin or calcium channel blockers if atrial fibrillation emerges
- In AF and mechanical valve → anti coagulants

Paroxysmal AF or permanent AF, LAT, prior embolism, spontaneous echocardiographic contrast in left atrium, large left atrium ( $\geq 60\text{ml/m}^2$ ) and Congestive HF

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- Given the increased risk of surgery, percutaneous mitral valvuloplasty is now the procedure of choice for pregnant patients with Rheumatic MS
  - Surgical commissurotomy has a lower maternal mortality rate, but is more risky for the fetus
  - In those where surgical commissurotomy is not appropriate, mitral valve replacement may be necessary
  - Surgical interventions should be reserved only for those patients who have symptoms refractory to medical therapy in whom valvuloplasty is contraindicated
  - One of the concerns with percutaneous mitral valvuloplasty → restenosis; requiring surgical intervention in the future

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- If a patient is diagnosed with MS early in pregnancy, the procedure should be delayed until 12 to 14 weeks to prevent radiation exposure during the period of organogenesis
  - If intervention becomes necessary after 20 weeks, it is best deferred to between 26 and 30 weeks gestation to prevent complications associated with births in the extremes of prematurity

Hameed A, Mehra A, Rahimtoola S. The role of catheter balloon for severe mitral stenosis in pregnancy. **Obstet Gynecol.** 2009; 114:1336–1360

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- Maternal mortality rates for percutaneous mitral valvuloplasty were 0.2%, and fetal mortality rates were 2%, which included elective terminations; overall, the procedural success rate was 98%
  - Lead shielding should be used & source should be away from the patient
  - Fetal loss is PMC → go upto 30%
  - Significant fetal mortality risk of 20% to 30% in MVR

# Antenatal management

Increased Left atrial pressure is the common cause for decompensation & Risk factors to be avoided

## GENERAL

- Referral to a hospital setting that represents an appropriate maternal level of care
- Low threshold for cardiac evaluation
- Moderate and high-risk CVD – multidisciplinary team
- Personalized approach plan – assess the risks – should be familiar to all

## SPECIFIC

- Frequency of visits depends upon the mWHO class
- Rate control, decongestive therapy & anticoagulants if necessary
- If intervention is needed – after 14 weeks ideal
- Stable heart disease – can safely go through pregnancy till term
- Maternal or fetal instability → need elective delivery

# Intrapartum management

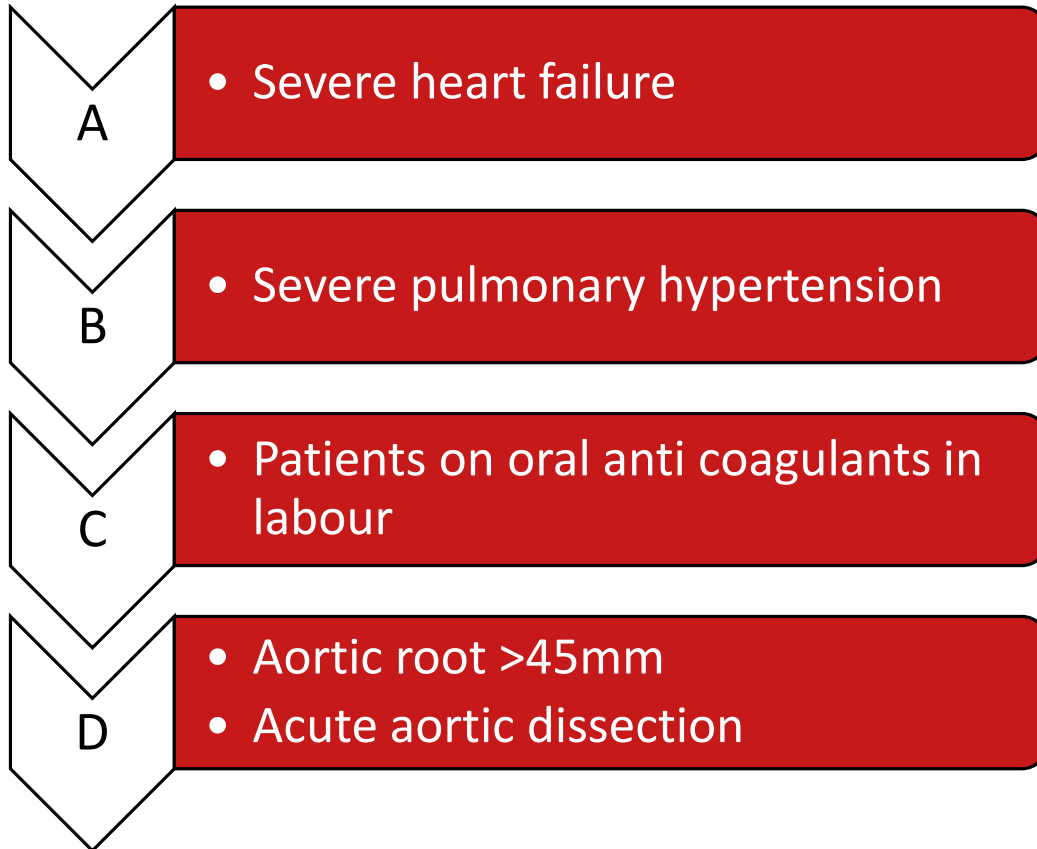
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- ❑ Usually CS is reserved for obstetric indications unless specific cardiac condition warrants
  - Most of the mWHO class IV → early elective delivery once fetal viability is achieved depends on their NICU set up
- ❑ Early epidural
- ❑ IE prophylaxis
- ❑ Encourage lateral decubitus
- ❑ Low dose oxytocin
- ❑ Avoid lithotomy / valsalva
- ❑ Cut short II stage
- ❑ Avoid methergin / Carboprost

## Carbetocin - Long acting oxytocin :

- Carbetocin 100mcg - can be given slowly as iv after dilution
- To be cautions in women with severe heart disease like ischemic heart disease, cardiomyopathy, valvular heart disease and heart failure

# Indications for caesarean section



## Mitral stenosis:

- Moderate to severe MS with Class III to IV symptoms
- Pulmonary HTN despite medical therapy
- In whom valvuloplasty could not be done



# Concerns during anaesthesia

Cardiac lesion	Preload	PVR	SVR	HR	Contractility
Atrial septal defect	↑	↑	↓	N	N
Ventricular septal defect (left→right)	↑	↑	↓	N	N
Ventricular septal defect (right→left)	N	↓	↑	N	N
Patent ductus arteriosus	↑	↑	↓	N	N
Idiopathic hypertrophic subaortic stenosis	↑	N	N↑	↓⊗	↓⊗
Coarctation	↑	N	↓	N	N
Pulmonary stenosis (valvular)	↑	↓	N	↓	↑
Pulmonary stenosis (infundibular)	↑	↓	N	↓	↓⊗
Aortic stenosis	↑	N	↑⊗	↓⊗	N↑
Mitral stenosis	N↓	N↓	N	↓⊗	N↑
Aortic regurgitation	N↑⊗	N	↓	N↑	N↑
Mitral regurgitation	N↑⊗	N	↓	N↑	N↑
Transposition of great arteries	N	N	N↑	↑	N
Tetralogy of Fallot	↑	N↓	↑	N↑⊗	N↓⊗

↑: Increase; ↓: Decrease; N: Normal; ⊗: Overriding consideration; PVR: Pulmonary vascular resistance; SVR: Systemic vascular resistance; HR: Heart rate

## Mitral stenosis:

- Under filled left ventricle
- Over pressured Right ventricle
- HR & ventricular Preload
- Measures to avoid anything increases PAP
- Graded Combined spinal epidural / EA is the choice – concern anticoagulants
- Severe cases NYHA III-IV / multi valvular → GA
- Special care – prosthetic valve on anticoagulants

Cardiac grid

# Post partum management

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- Close monitoring for first 24-48 hours
- Thromboprophylaxis
- Early post partum visit
- Psychological counselling
- Breast feeding

# Follow up

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- Post partum cardiac assessment – 6 / 12 weeks
- Drugs to be adjusted if necessary
- Extended follow-up depends on the severity
- Support
- Improve the cardiac status before the next planned pregnancy

# Contraception

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- IUCD
- Progestin only contraception – IUCD / Implant
- Barrier – failure rate
- Vasectomy / Tubectomy
- Combined OC Pill → TE is the concern
- DMPA – not a choice

## WHO CLASS



Maternal  
cardiac event  
rate

2.5–5%

Uncomplicated, small or mild PS; VSD; PDA; MVP  
with no more than trivial MR

Successfully repaired simple lesions (ASD or VSD,  
PDA, anomalous pulmonary venous drainage)

Isolated ventricular extra systoles & atrial ectopic  
beats

## WHO CLASS

II

Maternal  
cardiac event  
rate

5.7–10.5 %

Unoperated ASD or VSD

Repaired TOF

Most arrhythmias (supraventricular)

Turner without congenital heart disease

## WHO CLASS

### II-III

Maternal  
cardiac event  
rate

10-19 %

Mild ventricular impairment (EF >45%), heart transplantation, hypertrophic cardiomyopathy

Native or tissue valvular heart disease not considered WHO I or IV

Repaired Coarctation, Marfan syndrome without aortic dilatation, bicuspid valve with aorta <45 mm

Atrioventricular septal defect

## WHO CLASS

### III

Maternal  
cardiac event  
rate

19-27%

Mechanical valve

Moderate left ventricular dysfunction (EF 30-45%)

Moderate MS, Severe asymptomatic AS

Previous PPCM without residual dysfunction

Systemic right ventricle, Fontan circulation, unrepaired cyanotic heart disease, other complex CHD,

Marfan syndrome with aorta 40–45 mm

bicuspid aortic valve with aorta 45–50 mm



WHO CLASS  
IV

Maternal  
cardiac event  
40-100 %

Pulmonary hypertension

Eisenmenger syndrome

Systemic ventricular ejection fraction <30% or  
systemic ventricular dysfunction with NYHA class III–IV

Prior peripartum cardiomyopathy with any residual impairment of  
ventricular function

Severe MS, severe symptomatic AS

Marfan syndrome with aorta >45 mm, Vascular ED syndrome  
bicuspid aortic valve with aorta >50 mm, native severe coarctation

## References:

1. ACOG Practice bulletin No:212 May 2019
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4. High-Risk Cardiac Disease in Pregnancy Part I & II JOURNAL OF THE AMERICAN COLLEGE OF CARDIOLOGY 2016
5. Disease and Pregnancy Cardiol Ther 2017
6. Pregnancy Complicated by Valvular Heart Disease: An Update  
American heart Association 2014