Matters of the Heart: Cardiac Issues and Management in Pregnancy

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Introduction

- In developed countries, maternal heart disease (while very rare) is the leading cause of maternal death during pregnancy
- Hypertensive disorders are the most frequent CVS disorders encountered during pregnancy
- Congenital heart disease (CHD) is the next most common cause of CVS disorders in western countries (75 – 82% of non-hypertensive CVD presentations in pregnancy)
- Rheumatic valvular disease dominates in developing countries
- Women with cardiac disease have an increased risk of obstetric complications, including premature labour, pre-eclampsia and post-partum haemorrhage

Questions that need answers.

- How does any given medical condition affect the chances of successful pregnancy?
- How will pregnancy affect the medical condition?
- Are there any pregnancy issues specific to the condition?
- How will medication affect the baby?
- What kind of antenatal care should a woman have (and where) and can they have a natural delivery?
- Is the baby at increased risk of developing the condition?
- Are a patient's symptoms / signs due to 'normal pregnancy' or a deterioration in their medical condition? Or a new pregnancy-related complication?
- Who else needs to know about this patient?
- Are there any other factors that need to be taken into consideration?

Physiological adaptation in normal pregnancy

- Peripheral vasodilatation, mediated by endothelium-dependent factors (including NO synthesis upregulated by oestradiol and possibly vasodilatory prostaglandins)
- Produces a fall in systemic vascular resistance (SVR) and compensatory increase in cardiac output (CO) by 30-50%, predominantly achieved by an increase in stroke volume (SV) and a lesser increase in heart rate (HR).
- Increased SV is possible due to an increase in ventricular wall muscle mass and end-diastolic volume ie: the heart is dilated and contracts more strongly (‘rubber band analogy’)
- By 8/40, CO has increased by 20%

Physiological adaptation in pregnancy (2)

- Between 20/40 – 28/40, maximal cardiac CO is reached; there is a slight decline in SV towards term, but overall CO is mostly maintained by the increase in HR (10-20bpm)
- Pulmonary vascular resistance (PVR) decreases significantly in normal pregnancy; there is no significant change in central venous pressure or pulmonary capillary wedge pressure (PCWP).
- Decrease in serum colloid osmotic pressure gives approximately 30% reduction in colloid osmotic pressure / PCWP gradient, leading to an increased susceptibility of pregnant women to pulmonary oedema in situations of increased cardiac pre-load (eg fluid infusion) and / or increased pulmonary capillary permeability (eg pre-eclampsia)

A note on maternal positioning

- Close to term, pressure of the gravid uterus on the IVC will cause a reduction in venous return to the heart and a resultant decrease in SV and hence CO.
- There can be a reduction of up to 25% in CO with change from the lateral to the supine position, which has effects on placental (and fetal) blood flow
- Where possible, pregnant women should be nursed in the lateral position, or have the pelvis rotated to move the uterus off the IVC if she must be kept on her back.

Summary of changes (Nelson-Piercy, C Handbook of Obstetric Medicine, 1997)

Physiological Variable	Direction of change	Degree / timing of change
Cardiac output (CO)	↑	40%
Stroke volume (SV)	↑	
Heart rate (HR)	↑	10 – 20 bpm
Blood pressure	↓	First and second trimesters
	↑	Third trimester
Central venous pressure (CVP)	↔	
Pulmonary capillary wedge pressure (PCWP)	↔	
Systemic and pulmonary vascular resistance (SVR and PVR)	↓	25 – 30%
Serum colloid osmotic pressure	↓	10 – 15%

Intrapartum and postpartum

- CO increases in labour (up to 15% in first stage and 50% in second stage), both during and between contractions; uterine contractions will auto-transfuse 300-500mL of blood back into the circulation and the sympathetic response to pain and anxiety will further elevate both HR and BP
- Following delivery, there is an immediate rise in CO due to the relief of IVC obstruction and auto-transfusion of the utero-placental circulation back into systemic circulation. CO increases by 60-80%, followed by a rapid decline to pre-labour values within approximately an hour of delivery

Intrapartum and postpartum (2)

- Transfer of fluid from the extra-vascular space further increases venous return and SV
- Women with cardiovascular compromise are thus most at risk of APO during second stage and the immediate post-partum period
- By 2/52 post-partum, CO has nearly returned to normal pre-pregnancy values, though some haemodynamic changes (eg HTN due to pre-eclampsia) may take longer

- **Any condition which impairs the ability to make these adaptations has potentially serious implications for pregnancy**

So what is normal during pregnancy?

- On examination:
  - Bounding / collapsing pulse
  - Ejection systolic murmur – present in > 90% of pregnant women: may be quite loud and audible throughout the praecordium
  - Loud first heart sound and a third heart sound
  - Relative sinus tachycardia and ectopic beats
  - Peripheral oedema
- On ECG:
  - Atrial and ventricular ectopics
  - Q-waves (small) and inverted T-waves in III
  - ST segment depression and T-wave inversion in inferior and lateral leads
  - QRS leftward axis shift

Palpitations and ‘Dizziness’

- Very common symptoms in normal pregnancy
- Basic assessment:
  - Clinical history, including symptoms of GORD / exercise tolerance
  - BP and pulse / chest auscultation / examination of the peripheries
  - ECG
  - CBE / Fe studies / TFT’s / EUC / LFT’s incl Ca^{2+} and Mg^{2+}
- Caffeine reassessment
- Correction of iron deficiency +/- anaemia
- Always worth a 2 week trial of PPI (omeprazole / pantoprazole / esomeprazole)
- 24hr Holter and echo if concerning symptoms / duration – tends to be low-yield, but may provide reassurance. Should pair with symptom diary

POTS

- Very common diagnosis in this demographic and symptoms exacerbated by the normal physiological changes and relative deconditioning that comes with pregnancy
- Often associated with significant anxiety and functional debility
- (Fortunately) less commonly seen as part of a broader medical issue (eg Ehlers Danlos syndrome, SLE, other causes of autonomic neuropathy)
- POTS in isolation does not adversely affect pregnancy outcomes, and should not influence decisions around mode (and timing) of delivery
- Management remains similar to the non-pregnant population: exercise and diet with correction of exacerbating factors and attention to fluid, salt and fibre intake,
- Medication options – β-blockers / fludrocortisone are well described in pregnancy. Midadrine and ivabradine less so, but may be indicated, especially if concurrent conditions
- https://my.clevelandclinic.org/health/diseases/16560-postural-orthostatic-tachycardia-syndrome-pots

Arrhythmias (1)

- Identify precipitant / manage underlying cause
- More significant if occurs in the presence of underlying structural heart disease (eg mitral stenosis) or cardiomyopathy
- Treatment depends on accurate identification – ECG / Holter / implantable loop recorders & patient activated devices
- Paroxysmal supraventricular tachycardia (SVT) is the commonest arrhythmia encountered in pregnancy – usually diagnosed prior to pregnancy, but may become more symptomatic or frequent in pregnancy
- If previous successful ablation: likely to manage well, with nil significant issues

Arrhythmias (2)

- Management remains similar to non-pregnant:
  - Identify underlying trigger / exclude structural heart disease if appropriate
  - Valsalva manoeuvre (re-entrant SVT's)
  - Adenosine to terminate SVT's (or unmask atrial flutter) – doses of 6-12mg
  - B-blockers (**metoprolol** / **propranolol** / sotalol / atenolol): AF / WPW / prophylaxis of AF / SVT / VT / long QT syndromes
  - Verapamil – oral or IV (5-10mg gentle bolus) – second line for acute management of SVT
  - Digoxin can be used for rate control in AF if β-blockers are not tolerated
  - Flecainide is safe to use / continue if indicated
  - Lignocaine: stable VT
  - Cardioversion (with clexane for anticoagulation as appropriate)

Adult congenital heart disease (CHD)

- Commonest category of birth defects, with just under 1% of newborns affected: 50% will be women – management of these individuals both with and without surgery has significantly improved over the past few decades
- Increasing numbers are surviving into adulthood and are usually well enough to consider pregnancy – either because they have a lesion with good long-term prognosis or because they have had successful surgery
- Pre-pregnancy counselling and assessment is critical – prognostic factors associated with poor maternal outcomes have been prospectively evaluated and recognised

Known poor prognostic factors in CHD

- Poor maternal functional class (NYHA functional classification) – III / IV
  - I – no dyspnoea / uncompromised
  - II – dyspnoea on severe exertion / slightly compromised
  - III – dysnoea on mild exertion / moderately compromised
  - IV – dyspnoea at rest / severely compromised
- Cyanosis / History of TIA / Heart failure / Left-sided arrhythmia / Left heart outflow obstruction / Impaired ventricular function
- Maternal cyanosis in CHD (excluding Pul HTN) is also associated with fetal and neonatal complications:
  - Sats > 90% - LBR 92%
  - Sats ≥ 85 – 90% - LBR 63%
  - Sats < 85% - LBR 12%

Other factors to take into account:

- Thromboembolic risk and need for anticoagulation (eg Fontan's repair)
- Vaginal delivery is generally the safest option for the mother with reduction in both blood loss and VTE risk, as well as less abrupt haemodynamic changes (especially when assisted by early effective analgesia and short second stage). Lack of predictable delivery timing is the only potential downside
- Some absolute indications for LSCS include:
  - Aggressive aortic pathology (eg Marfan's with aortic root diameter > 45mm, aortic aneurysm)
  - Acute intractable heart failure
  - Severe pulmonary HTN (including Eisenmonger's)
  - Active anticoagulation with warfarin

Still more things to consider:

- The risk of CHD is increased in the offspring of parents with CHD – 6% if the mother had CHD, 2% if the father does (higher if both parents are affected)
- All women with CHD (or other possible genetic cardiac disease) should be offered a fetal echo between 19/40 - 22/40 at a specialised centre; further antenatal echo evaluation may be indicated.
- Referral for pre-conception counselling as early as possible is ideal – if not, referral ASAP in pregnancy and liaison with appropriate services
- Phone a friend!

Cardiomyopathies -HCM

- Hypertrophic cardiomyopathy – approximately 70% of cases are familial, with autosomal dominant inheritance
- Previously regarded as rare and associated with high risk of sudden death, better (and more frequent) screening has shown it to be more common, and often benign
- β-blocker prophylaxis should be continued (or commenced) for symptomatic women
- Generally well-tolerated in pregnancy
- Care required with regional analgesia and peripartum fluid management to avoid hypotension and hypovolaemia, which can exacerbate left ventricular outflow tract obstruction

Cardiomyopathies - PPCM

- Peripartum cardiomyopathy – development of heart failure (LVEF <45% / LVEDP > 2.7cm/m²) in the absence of a known cause and without prior heart disease prior to the last month of pregnancy and up to and including the fifth month post-partum. Most common in the first month post-partum
- <u>Management:</u>
  - Elective delivery if antenatal
  - Thromboprophylaxis: 25-40% rate of systemic embolisation and 5% risk of ischaemic stroke untreated.
  - Usual management for HF: frusemide / hydralazine & nitrates / carvedilol / bisoprolol / digoxin / ionotropes. Can add ACE-inhibitors post-delivery
  - IABP and LVAD's, with transplantation in severe cases unresponsive to conventional treatment

Cardiomyopathies: PPCM

- Prognosis and recurrence
- improved mortality and morbidity compared to historical controls
- 50% will make full recovery, but will still have up to 25% risk of recurrence in future pregnancy: stress echo (dobutamine or exercise) may identify impaired contractile reserve in women with “normal” LVEF pre-conception
- 50% risk of worsening heart failure and 25% maternal mortality rate in subsequent pregnancy in women who do not return to baseline LV dimensions and function
- Contraception with progesterone-only devices (ie mirena / Kyleena / Implanon) is appropriate – avoid oestrogen-containing preparations

Cardiomyopathies – DCM

- Other causes of dilated cardiomyopathy include:
  - Infections: viral / bacterial / fungal / parasitic
  - Neuromuscular – muscular dystrophy
  - Nutritional deficiencies – niacin, thiamine, selenium
  - CT diseases – SLE / Rheumatoid arthritis
  - Vascular – Kawasaki disease
  - Haematological – thalassaemia / sickle cell disease / iron deficiency anaemia
  - Drugs – alcohol / iron overload
  - Endocrine – hypo and hyperthyroidism / hypoparathyroidism / phaeochromocytoma
  - Metabolic – haemochromatosis / glycogen storage disorders
- Pregnancy contraindicated if LVEF < 30% or NYHA classification III or IV

Myocardial infarction / acute coronary syndromes

- Rare in women of child-bearing age, but becoming more frequent as women delay childbirth and acquire more cardiovascular risk factors - smoking remains a significant contributor to maternal IHD
- Acute MI / ACS occurs most commonly in third trimester, peripartum and postpartum – often without a history of angina. Arterial dissection is particularly associated with the postpartum period
- LAD territory and anterior wall of the left ventricle are the most commonly affected sites
- Troponins I and T are not altered in normal pregnancy, but Trop I is increased in pre-eclampsia, pulmonary embolism, atrial fibrillation and myocarditis (unlike Trop T)

Myocardial infarction / acute coronary syndromes - continued

- Coronary angiogram, percutaneous coronary interventions and stenting should be performed in pregnancy if indicated – bare metal stents may be preferred to drug-eluting stents due to the shorter required duration of dual anti-platelet therapy – both aspirin and clopidogrel should be used if indicated
- Angioplasty carries a higher risk of coronary artery dissection in the pregnant population, but should be performed if indicated
- CABG may be required in an emergency and should not be withheld – maternal outcomes are generally good, and comparable to the non-pregnant population. Fetal mortality is high (30-40%).

Aortopathies

- Increased risk of both aneurysm formation and aortic dissection – rare, but associated with very high mortality – again, most dissections occur in third trimester or postpartum
- Most deaths occur in women not known to have aortopathy, but post-mortem DNA analysis should be performed, as most will have hereditary aortopathy and family screening should be offered
- Hereditary Thoracic Aortic diseases (HTAD's) include syndromes such as Marfan's, Loeys-Dietz and Vascular Ehlers-Danlos, as well as non-syndromal aortic aneurysms.
- Other congenital lesions associated with aortic pathology include bicuspid aortic valve, tetralogy of Fallot and co-arctation of the aorta, Turner's syndrome can also be associated with aortic pathology.
- Pre-pregnancy imaging of the entire aorta is indicated in women with known or hereditary aortic disease – CT or MRI – and calculations of diameter should take BSA into account
- All women with aortic disease should be counselled about the risk of aortic dissection

Aortopathies - continued

- Parity appears to be associated with increased aortic diameter
- Strict blood pressure control (<120/80) is indicated during pregnancy, and β-blockers are preferred
- Regular antenatal echos should be performed on women with dilation of the ascending aorta; MRI without gadolinium can be used to monitor women with disease in the descending aorta
- If ascending aorta < 40mm diameter – NVD is recommended
- Between 40-45mm – LSCS or NVD with early epidural and managed second stage is advised; beyond 45mm – LSCS.

mWHO Risk Class I

- No detectable increased risk of maternal mortality and no or mild increase in morbidity: 2-5% risk of maternal cardiac event
- Management:
  - Pre-pregnancy / pregnancy counselling
  - Care, including delivery, at local hospital
  - Cardiology evaluation 1-2x/ pregnancy
- Uncomplicated or mild:
  - Pulmonary stenosis
  - Patent ductus arteriosis (PDA)
  - Mitral valve prolapse
- Successfully repaired simple lesions (atrial or ventricular septal defect, PDA, anomalous pulmonary venous drainage)
- Atrial or ventricular ectopic beats (isolated)

mWHO Risk Class II

- Small increased risk of maternal mortality or moderate increase in morbidity: 6-10% maternal cardiac event rate
- Management:
  - Pre-pregnancy / pregnancy counselling
  - Specialist consultation / counselling (Cardiology, Obs Med / MFM / Anaesthetics)
  - Care, including delivery, at local hospital
  - Cardiology evaluation every trimester
- Unoperated atrial or ventricular septal defects
- Mild aortic stenosis
- Repaired Tetralogy of Fallot (TOF) or aortic coarctation
- Most arrhythmias (supraventricular arrhythmias)
- Turner’s syndrome without congenital cardiac disease

mWHO Risk Class II / III

- Intermediate increased risk of maternal mortality or moderate to severe increase in morbidity: 11-19% maternal cardiac event rate
- Management:
  - Pre-pregnancy / pregnancy counselling
  - Specialist consultation / counselling (Cardiology, Obs Med / MFM / Anaesthetics)
  - Care, including delivery, at appropriate level hospital
  - Cardiology evaluation every trimester
- Mild left ventricular impairment (EF > 45%)
- Hypertrophic cardiomyopathy

mWHO Risk Class II / III - continued

- Native or bioprosthetic valve disease not considered mWHO Risk Class I or IV (mild mitral stenosis or moderate aortic stenosis)
- Marfan or other HTAD syndrome without aortic dilation
- Aorta < 45mm in bicuspid aortic valve pathology
- Repaired coarctation without residua (non-Turner's)
- Atrioventricular septal defect

mWHO Risk Class III

- Significantly increased risk of maternal mortality or severe morbidity: 20-27% maternal cardiac event rate
- Management:
  - Pre-pregnancy / pregnancy counselling
  - Specialist consultation / counselling (Cardiology, Obs Med / MFM / Anaesthetics)
  - Care, including delivery, at appropriate level hospital
  - Cardiology evaluation every 1-2 months
- Moderate left ventricular impairment (EF 30-45%)
- Previous peripartum cardiomyopathy without any residual left ventricular impairment

mWHO Risk Class III - continued

- Mechanical valve
- Systemic right ventricle with good or mildly decreased right ventricular function
- Uncomplicated Fontan circulation
- Unrepaired cyanotic heart disease
- Other complex congenital heart disease
- Moderate mitral stenosis
- Severe asymptomatic aortic stenosis
- Moderate aortic dilation (40-45mm in Marfan syndrome or other HTAD, 45-50mm in bicuspid aortic valve, Turner syndrome with aortic size index 20-25mm/m^2, Tetralogy of Fallot <50mm)

mWHO Risk Class IV

- Pregnancy contraindicated: discuss TOP
- Extremely high risk of maternal mortality or severe morbidity: >27% maternal cardiac event rate
- Management:
  - Specialist consultation / counselling (Cardiology, Obs Med / MFM / Anaesthetics)
  - Care and delivery at appropriate level hospital: may require prolonged antenatal admission
  - Cardiology evaluation every month (minimum)
- Pulmonary arterial hypertension
- Severe systemic ventricular dysfunction (EF < 30%, NYHA III- IV)
- Previous peripartum cardiomyopathy with any residual left ventricular dysfunction

mWHO Risk Class IV - continued

- Severe mitral stenosis
- Severe symptomatic aortic stenosis
- Systemic right ventricle with moderate to severely decreased right ventricular function
- Severe aortic dilation (>45mm in Marfan syndrome or other HTAD, >50mm in bicuspid aortic valve, Turner syndrome with aortic size index >25mm/m², Tetralogy of Fallot > 50mm)
- Vascular Ehlers-Danlos (Type IV)
- Severe (re)coarctation
- Fontan circulation with any complication

Specific considerations for resuscitation in pregnant women

- In addition to the usually causes of cardiac arrest, some pregnancy and post-partum-specific causes to consider:
  - Amniotic fluid embolism
  - Pulmonary embolism
  - Peripartum cardiomyopathy
  - Acute coronary / aortic dissection
- Compressions : breaths ratio – 30:2 – ‘Staying Alive’
- More difficult to get effective CPR due to:
  - Aortocaval compression
  - Enlarged breasts
  - Diaphragmatic splinting
- Wedge RHS of patient / raise right hip / manually displace uterus to LHS
- Early intubation to prevent aspiration (delayed gastric emptying in pregnancy)
- Emergency LSCS after 5 mins CPR recommended as next line

Contraception and family planning

- In general, in any woman where there is an increased risk of VTE or HTN is likely to either be exacerbated or cause additional morbidity, the combined oral contraceptive pill is contraindicated.
- Progesterone-only devices, including IUD's are recommended; as there is a risk of vagal response and hypotension with insertion of larger IUD's, it is best performed within an appropriate hospital setting in some women (eg with Fontan repairs or Eisenmonger's syndrome)
- Barrier methods are unreliable, but safe.
- Sterilisation may be considered, but should be performed in an appropriately-equipped and staffed centre; some conditions may warrant hysteroscopic tubal ligation under regional block (eg pul HT, cyanotic women)
- Termination of pregnancy by either medical or surgical means generally has similar effectiveness and safety profiles as in the general population, but surgical termination may be preferred due to the increased chance of requiring unanticipated operative evacuation following medical termination (2.1% cf 0.6%) – again, in women with high-risk lesions, procedures should be undertaken in appropriate hospitals

Final words

- Pregnant women with significant cardiac disease should generally undergo the same processes of investigation and management as non-pregnant patients – untreated, maternal cardiac compromise carries a carries a high risk of both maternal and fetal mortality and morbidity
- “In the case of an emergency, drugs that are not recommended by the pharmaceutical industry during pregnancy and breastfeeding should not be withheld from the mother. The potential risk of a drug and the possible benefit of the therapy must be weighed against each other.”

(Almost)

- Cardiac medication options include: metoprolol, carvedilol, verapamil, diltiazem, digoxin, warfarin, LMWH, aspirin, frusemide, flecainide, adenosine.
- Cardioversion can be safely performed in pregnancy.
- Plan location of antenatal care and delivery – ideally by 30/40; may need delivery in a centre where specialised cardiac HDU and anaesthetics services are available; many women can have safe NVD.
- Careful communication between GP, Cardiologist, Obstetrician, Anaesthetist and Obstetric Physician is ideal

Other resources:

- WCH
  - on-call Physician
  - Drug Information service 8161 7555 – Mon – Fri, 9 - 5
- SA PPG's: Cardiac Disease in Pregnancy
- 2018 ESC Guidelines for the management of cardiovascular disease during pregnancy. Published in the European Heart Journal 2018; **39**, 3165 – 3241.