Cardiac Disease in Pregnancy

Key Points

- Pre-pregnancy counselling is essential for all women of childbearing age with known (or previously repaired) cardiac disease
- All women should be booked under the care of a consultant with a specialist interest in Maternal Medicine
- Multi-disciplinary care is essential
- Pregnancies should be managed according to the mWHO and NYHA classification

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Abbreviations

LQTS	**Long QT Syndrome**
MMN	Maternal Medicine Network
mWHO	Modified WHO (World Health Organisation)
NYHA	New York Heart Association
OAC	Oral anticoagulant
PAH	Pulmonary Arterial Hypertension
VTE	Venous thromboembolism

1. Introduction

The MBRRACE report from November 2024 found that cardiac disease was the second most common cause of maternal death in the UK between 2020-2024. In the 2017-2019 MBRRACE report, cardiac disease was the leading cause of death in women. 77% of the women who died from cardiac disease were not known to have pre-existing cardiac disease. There appear to be several factors contributing to this.

There are increasing numbers of pregnant women of advanced maternal age, and these are more frequently associated with cardiovascular risk factors (diabetes, hypertension, obesity). A larger proportion of women with congenital heart disease are now reaching child-bearing age. It is recommended that these women receive pre-pregnancy counselling and that their management during pregnancy and delivery is overseen by a multidisciplinary team (cardiologist, obstetrician and anaesthetist) that specialise in high-risk pregnancies. An increasing number of migrant women, rheumatic heart disease in pregnancy has re-emerged. Cardiac symptoms may also be missed or under-investigated in pregnancy or in the early post-partum period. This may be due to a lack of thorough history/examination, reluctance to perform certain diagnostic tests on a pregnant woman, or simply not considering cardiac disease as a potential diagnosis. A high index of suspicion is required and early involvement of senior clinicians from the obstetric and cardiology multidisciplinary team is vital. Once a diagnosis is made, risk stratification should occur, and the woman might need transfer of care if her cardiac condition is moderate or severe (WHO II – IV).

1.1 Normal physiological cardiopulmonary changes in pregnancy

- Systemic and pulmonary vascular resistance decrease in pregnancy
- Blood pressure decreases to a nadir in the second trimester, then increases in the third trimester, returning close to preconception levels postpartum
- Cardiac output and plasma volume reach 40-50% above baseline by 32 weeks gestation
- Increase in cardiac output is due to raised stroke volume in the first half of pregnancy, followed by a raised heart rate thereafter
- Maximum cardiac output is during labour and immediately postpartum (60-80% above levels prior to labour). This is due to uterine contractions causing an extra 300-500mls of blood to be released into the circulation during labour, and relief of IVC obstruction/contraction of the uterus causing extra blood to enter the circulation postdelivery.
- Due to the above physiological changes, aortic stenosis and mitral stenosis are poorly tolerated in pregnancy. Both these conditions are associated with a reduced capacity to increase cardiac output. On the other hand, aortic regurgitation and mitral regurgitation are well tolerated- owing to the fall in peripheral vascular resistance during pregnancy and its effect to reduce afterload. Women with cardiac disease are most at risk of pulmonary oedema during the second stage of labour and immediately postpartum.

2. Antenatal Care

• Women with a history of maternal cardiac disease should be booked < / = 12/40 into a maternal medicine obstetric clinic. They carry an increased risk of premature labour, pre-eclampsia and post-partum haemorrhage.

• The booking midwife should encourage the woman to gather as much historical medical information regarding her condition prior to her obstetric appointment.

• Aspirin 150mg orally to be taken at night should be recommended from 12 weeks' gestation until birth if active cardiac disease present.

• Women should be referred to be seen by an obstetric anaesthetist. Ideally this review should take place prior to 30 weeks' gestation of pregnancy.

• The risk of complications is largely dependent upon the underlying cardiac diagnosis and the presence of additional co-morbidities. Disease specific risk should be assessed using the modified World Health Organisation (mWHO) classification. See Appendix 1.

• Women that are classified as moderate or high risk (mWHO II-III, III, and IV) should be managed in a specialist, tertiary centre. However, care must be individualised and it may be that women in mWHO I and II may also need tertiary level input as guided by the MDT.

• Women that are classified as mWHO IV would be advised not to fall pregnant.

• According to their cardiac diagnosis, it is advisable to discuss the woman's antenatal and intra-partum care in a MDT maternal medicine meeting (either local or tertiary centre led within Maternal Medicine Network (MMN)). This should ideally occur between 18- 30 weeks' gestation.

• Maternal cardiac disease alone is not an indication for serial fetal growth scans in the third trimester. Medication history and additional risk factors may indicate that these are necessary.

• Women with a pre-existing cardiac problem should be managed according to their antenatal and intrapartum care plan and involving the relevant physicians and anaesthetists. Multi-disciplinary care and liaison between senior clinicians is key in their management.

• Contraceptive advice should be re-iterated to woman and a robust plan of care put in place postnatally.

2.1 Screening of the fetus

For a fetus whose parents do not have congenital heart disease, the background risk is 1%. Heritability from parents varies between 3% and 50%, depending on the type of disease. Those whose parents have an autosomal dominant condition such as Marfan syndrome or long QT syndrome, will have an inheritance risk of 50%. However, most congenital heart disease is polygenic and therefore the risk is harder to define. Genetic testing and counselling may be useful in those women who have a history of venous thromboembolism (VTE) and/or pulmonary arterial hypertension (PAH), Long QT syndrome (LQTS) or other channelopathies, thoracic aortic pathology and congenital heart disease with genetic abnormalities. It is recommended that women with inherited cardiac conditions should be referred to the genetics team as the baby will require follow up.

Nuchal fold thickness at the 12-week scan has a sensitivity of 85% and specificity of 99% for major congenital heart disease. The incidence of major congenital heart disease with normal nuchal fold thickness is approximately 1/1000.

All women with congenital heart disease should be offered a fetal echocardiogram between 19-22 weeks gestation. Approximately, 50% of all congenital cardiac malformations will be identified. Please refer the woman via the Antenatal Screening Midwives. Women who are booked at FPH will be referred to Evelina Children's Hospital at Guy's and St Thomas' Hospital. Women who are booked at WPH will be referred to Fetal Medicine Unit, John Radcliff Hospital, Oxford. Offspring complications occur in 18–30% of patients with heart disease, with neonatal mortality between 1–4%

If there is suspicion or confirmation of a fetal cardiac defect, delivery is likely to be advised at a tertiary neonatal unit.

2.2 Timing and mode of delivery

An individualised birth plan should be formulated by the consultant obstetrician and midwife, in conjunction with a cardiologist and anaesthetist. This should cover all three stages of labour plus the postpartum period and be accessible in the patients' notes. In general, caesarean section should be advised for obstetric indications only, unless:

- Significant aortic dilatation is present
- NYHA III or IV heart disease
- Pulmonary arterial hypertension (incl. Eisenmenger's syndrome)
- Woman is taking oral anti-coagulants (OACS)

Induction of labour should be considered at 40 weeks' gestation in all women with cardiac disease, unless otherwise documented.

2.2.1 Premature Labour

Tocolytics should not be commenced without prior discussion with a consultant obstetrician, as they may severely compromise cardiac function. Atosiban has the least cardiovascular side effects of all the tocolytics and is therefore the tocolytic of choice for women with severe cardiac disease. Steroids for fetal lung maturity are not contraindicated. Magnesium sulphate infusions for neuroprotection can also be used but may cause hypotension, so should be used with care and strict adherence to frequency of blood pressure monitoring.

3. Intrapartum Management

- The documented delivery care plan should be documented in the patients EPIC notes and accessible to all, 24 hrs/day. The plan must be adhered to, and cardiology/ medical input sought if concerns arise.

- Women should be advised to birth on the labour ward.
- On admission, a multi-disciplinary team review (including midwife, consultant obstetrician and anaesthetist +/- physician) should occur. A routine examination including baseline observations and heart sounds and lung fields auscultation should be performed.
- A 16-gauge (grey cannula) should be inserted once in established labour.
- A fluid balance chart should be commenced upon arrival to the maternity unit.
- For many cardiac conditions, epidural analgesia is beneficial due to pain relief and subsequent reduction in circulating catecholamines that may be detrimental to cardiac function.

Management of 1st and 2nd stage of labour

Women with mWHO I or II disease can be offered the same analgesia and anaesthesia as low risk women. Women with mWHO I or NYHA I disease can receive standard management of fluid balance during the intrapartum period.

Induction of labour using Propess®, Prostin® or misoprostol can be safely used in a woman with cardiac disease. Mechanical methods may be preferable when a drop in systemic vascular resistance would be detrimental to the woman. Amniotomy and oxytocin infusion is safe, unless otherwise documented.

Women with mWHO I or II disease may be allowed up to 2 hours for the passive second stage to allow descent of the fetal head and minimise time spent in the active phase of labour. The woman should be reviewed by an obstetrician after 1 hour of pushing, with a view to expediting delivery. Women with mWHO III or IV disease may benefit from elective instrumental delivery.

Management of 3rd Stage of labour

In general, ergometrine/ syntometrine are often contraindicated as they cause a hypertensive surge. A syntocinon bolus can cause tachycardia and hypotension. For women with moderate or severe cardiac conditions, syntocinon should be given as a slow bolus intravenously (Syntocinon 5 units in 20 ml of Sodium Chloride 0.9% over 20 mins via an infusion pump). In cases where there is uncertainty about what to use for the third stage of labour, then this regime should be used as it has the least vasoactive side effects.

In selected cases, Women with mWHO I or II disease may have the third stage of labour managed as per low-risk women in trust guidelines (i.e. they do not need to have active management).

Women with mWHO III or IV disease should be given oxytocin as the uterotonic of choice, followed by Misoprostol pr as second line. Carboprost can be considered when there is significant blood loss secondary to atony. Ergometrine should be avoided. Generally, women should be assisted to sit up as soon as possible after delivery.

4. Postpartum management

- Multi-disciplinary care plan should be followed.
- Careful attention must be paid to woman's fluid balance for a minimum of 24 hours, possibly longer depending upon cardiac condition. Please refer to individual postnatal care plans. This should be documented on a fluid balance chart.
- Care should be delivered on the labour ward for a minimum of 24 hours, where possible.
- Observations should be performed every 30 minutes for 4 hours and then every 60 minutes for a further 12 hours for those with complex cardiac disease. Please follow individual postnatal care plans.
- Uterine hypotonia: Syntocinon infusions can be used although in some women the volume of fluid may need to be limited (ie a more concentrated infusion). Mechanical methods such as bimanual compression, B Lynch suture and Bakri balloon can also be used in problematic hypotonia. Misoprostol (1000 micrograms rectally) should be used in preference to carboprost (Haemobate) since the former is less vasoactive. If a woman is becoming unstable because of blood loss due to haemorrhage from uterine hypotonia then syntometrine can be used (in this situation the hypertensive surge won't be a problem).
- Timing of discontinuation of epidural and commencement of thromboprophylaxis should be clearly documented in liaison with the anaesthetist.
- Early MDT involvement of the Cardiologist/ Physician, Anaesthetist and Obstetric consultant in the presence of tachycardia, reduced saturation, tachypnoea, chest pain or concerns with fluid balance.
- Contraceptive advice should be re-iterated to woman and plan of care put in place.

Sources of Support

Women should be made aware of sources of support. Women may wish to access support at any stage from the time when they are first considering pregnancy until after the baby is born. Women with heart disease may have difficult decisions to make and may not always have a successful outcome to the pregnancy. Women may find access to peer support helpful (i.e. British Heart Foundation (BHF) or the Grown Up Congenital Heart (GUCH) Patient's Association and should be given contact details (see Appendix 2).

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Appendix 1: Modified WHO (mWHO) classification of maternal cardiovascular risk

All women should be risk assessed using the mWHO risk classification. Those that are classified as moderate or high risk (mWHO II-III, III, and IV) should be managed in a specialist centre. Those that are classified as mWHO IV would be advised not to fall pregnant.

mWHO I

· No increased risk maternal mortality with mild increased risk in morbidity

· 2.5-5% maternal cardiac event rate

· Can have antenatal care at local hospital

· Can deliver at local hospital

mWHO II

· Small increased risk maternal mortality with moderate increased risk in morbidity

· 5.7- 10.5% maternal cardiac event rate

· Can have antenatal care at local hospital

· Can deliver at local hospital

mWHO II-III

· Intermediate increased risk maternal mortality with moderate-severe risk in morbidity

· 10-19% maternal cardiac event rate

· Antenatal care at tertiary centre

· Delivery at tertiary centre

mWHO III

· Significantly increased risk maternal mortality or severe morbidity

· 19-27% maternal cardiac event rate

· Antenatal care at tertiary centre

· Delivery at tertiary centre

mWHO IV

· Extremely high risk maternal mortality or severe morbidity

· If pregnancy occurs, termination should be discussed

· 40-100% maternal cardiac event rate

· Antenatal care at tertiary centre

· Delivery at tertiary centre In cases of heart failure, the New York Heart Association (NYHA) classification should be used.

· NYHA Class I: No limitation of physical activity

· NYHA Class 2: Comfortable at rest, but ordinary physical activity causes symptoms

· NYHA Class 3: Comfortable at rest, but less than ordinary physical activity causes symptoms

· NYHA Class 4: Symptoms at rest, and unable to carry out any physical activity without discomfort Poor pregnancy outcome is more likely in NYHA Class 3 or Class 4, regardless of the specific lesion.

Table 1. mWHO risk classification

mWHO I	mWHO II	mWHO II-III	mWHO III	mWHO IV
Small or mild: - Pulmonary stenosis - PDA -mitral valve prolapse	Unoperated ASD/VSD	Mild left ventricular impairment (ejection fraction > 45%)	Moderate left ventricular impairment (ejection fraction 30-45%)	Severe systemic ventricular dysfunction (ejection fraction < 30%)
Successfully repaired simple lesions: - ASD/VSD -PDA - Anolamous venous drainage	Repaired Tetralogy of Fallot	Hypertrophic cardiomyopathy	Mechanical valve	Pulmonary arterial hypertension
Atrial or ventricular ectopic beats	Supraventricular arrythmias	Native or tissue valve disease not considered I or IV (e.g., Mild mitral stenosis, Moderate aortic stenosis)	Previous peripartum cardiomyopathy without residual impairment	Previous peripartum cardiomyopathy with any residual impairment
	Turners Syndrome without aortic dilatation	Marfans without aortic dilatation	Moderate mitral stenosis/Severe asymptomatic aortic stenosis	Severe mitral stenosis/Severe symptomatic aortic stenosis
		Bicuspid aortic valve pathology with aorta < 45mm	-Marfans with aortic dilatation 40-45mm -Biscupid aortic valve/ Turners/Tetralogy with aortic dilatation 45-50mm	-Marfans with aortic dilatation >45mm -Bicuspid aortic valve/Turners/Tetralogy with aortic dilatation >50mm
		Repaired coarctation	Ventricular tachycardia	Vascular Ehlers Danlos
		Atrioventricular septal defect	Unrepaired cyanotic heart disease	Severe coarctation
			Systemic right ventricle with good ventricular/mildly impaired function	Systemic right ventricle with moderate/severely impaired ventricular function
			Fontan circulation	Fontan with any complication

Appendix 2: Patient Support Groups for women with Cardiac Disease

British Heart Foundation

Website: www.bhf.org.uk

Grown Up Congenital Heart Disease (GUCH) Patients Association

Website: www.guch.org.uk

Full version control record