Aneurin Bevan University Health Board

Management of Cardiac Disease in Pregnancy

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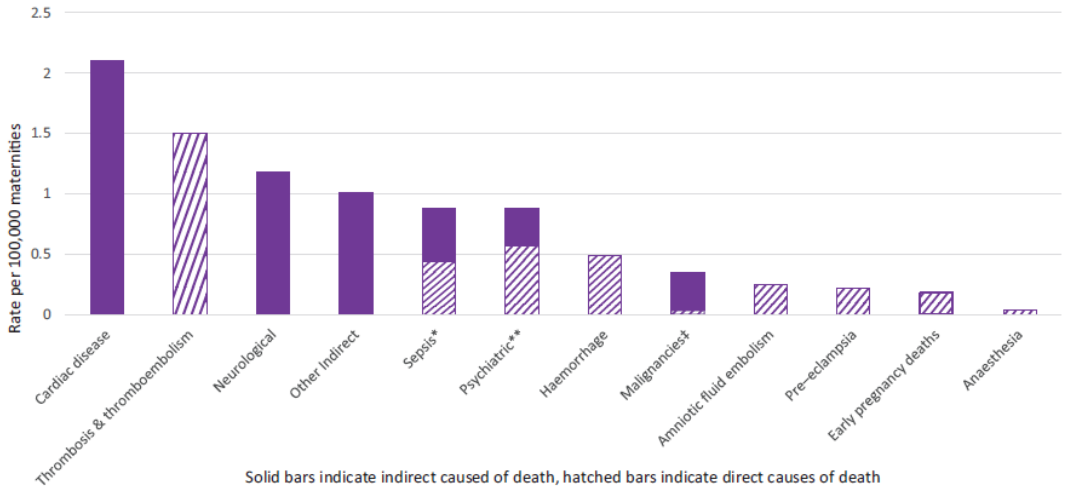
Contents:

Introduction

This policy aims to cover management of women with cardiac disease. Cardiac disease remains the largest single cause of indirect maternal deaths. There has been no change in the maternal mortality rate from cardiac disease since enhanced case ascertainment was introduced (RR 0.90, 95% CI 0.61-1.36 when comparing 2015-17 with 2003-05). The maternal mortality rate from cardiac disease in the UK for 2015–17 is 2.1 per 100,000 maternities. Overall, a quarter of women (24%) died from ischaemic causes, and almost one third from myocardial disease/cardiomyopathy (27%). Although there has been an apparent decrease in the proportion of women dying from sudden arrhythmic cardiac deaths with a morphologically normal heart (SADS/MNH) amongst those dying from cardiovascular causes, there is no statistically significant decrease in the mortality rate from SADS/MNH between 2009-14 and 2015-17.

The risk of cardiovascular disease in pregnancy has increased due to several factors such as advancing maternal age in pregnancy, increased prevalence of diabetes/ hypertension/ obesity/ smoking, advances in treatment of congenital heart disease resulting in increased number of women reaching adulthood and increased immigrant population with underlying cardiac disease.

Figure 2.3: Maternal mortality by cause 2015-17



Hatched bars show direct causes of death, solid bars indicate indirect causes of death;
*Rate for direct sepsis (genital tract sepsis and other pregnancy related infections) is shown in hatched and rate for indirect sepsis (influenza, pneumonia, others) in solid bar
**Rate for suicides (direct) is shown in hatched and rate for indirect psychiatric causes (drugs/alcohol) in solid bar
‡Rate for direct malignancies (choriocarcinoma) shown in hatched and rate for indirect malignancies (breast/ovary/cervix) in solid bar
Source: MBRRACE-UK

Policy Statement

This document is a procedure designed to support safe and effective practice*.*

Aims

To provide support for clinical decision making.

Scope

This guideline applies to all clinicians working within maternity services.

Roles and Responsibilities

The Maternity Management team.

Monitoring and Effectiveness

Local service Improvement Plan will guide monitoring and effectiveness.

Equality

This policy has undergone an equality impact assessment screening process using the toolkit designed by the NHS Centre Equality & Human Rights. Details of the screening process for this policy are available from the policy owner.

Appendices

Key Abbreviations:
NYHA – New York Heart Association ECHO - Echocardiography ACHD – Adult Congenital Heart Disease MVA – Mitral Valve Area AVA – Aortic Valve Area SVT – Supraventricular Tachycardia LVEF – Left Ventricular Ejection Fraction LVOT – Left ventricular Outflow Tachycardia CHD – Congenital Heart Disease AS – Aortic Stenosis MS – Mitral Stenosis PS – Pulmonary Stenosis AR – Aortic Regurgitation MR – Mitral Regurgitation TR – Tricuspid Regurgitation TOF – Tetrology of Fallot COA – Coarctation of Aorta ASD – Atrial Septal Defect VSD – Ventricular Septal Defect PDA – Patent Ductus Arteriosus MI – Myocardial Infarction HOCM – Hypertrophic Obstructive Cardiomyopathy EFM – Electronic Fetal Monitoring ECG - Electrocardiography HDU – High Dependancy Unit MEOWS – Modified Early Obstetric Warning Score CWS- Central Work Station ICD – Implantable Cardioverter-defibrillator AVRT / AVNRT – Atrioventricular Re-entrant Tachycardia, AV Node re-entrant Tachycardia WPW – Wolff-Parkinson-White Syndrome AF- Atrial Fibrillation

Cardiovascular Physiology in Pregnancy:

- Increase in cardiac output 30-50 %
- Increase in Plasma volume 40%
- Increase in heart rate 15%
- Increase in Stroke Volume 30%
- Blood Pressure – Systolic constant, Diastolic 15% decrease
- Reduction in systemic vascular resistance 25%
- Increased hypercoagulability of blood
- Cardiac output increases further 25% in 1st stage, 50% when pushing, 80% in early postpartum phase

Poor predictors of Cardiac events:

- Prior congestive cardiac failure/ stroke or arrhythmia
- Baseline NYHA > II, or cyanosis
- Left heart obstruction – MVA <2cm2 or AVA <3 cm2
- Left ventricular outflow tract gradient >30 mmhg on ECHO
- Left ventricular ejection fraction < 40%

Risk of Maternal mortality based on the Cardiac lesion

Low risk (1-2%)	Moderate Risk (2 -10%)	High Risk (10-50%)
• Corrected CHD (ASD,VSD,PDA) • Small ASD,VSD,PDA without shunts • Repaired COA • Mild MS/AS/PS • Mild – mod MR/AR/TR • SVTs/pacemakers without major underlying cardiac lesion, but seen by Local Cardiology	• Mod AS/MS/PS • Partially corrected TOF, Mustard/Senning surgeries • Deteriorating NYHA class • LVEF<40% • Severe AR/MR/PR • Marfan's with aortic root <4cm • Previous MI/ACS	• Severe AS/MS/PS • Native COA • PHT including Eisenmenger's) • Cyanotic lesions • Severe LVOT obstruction • Marfan's (aortic root>4cm) • LVEF <30% • h/o peri-partum cardiomyopathy • Ebstein's anomaly • HOCM • Type IV Ehlers Danlos • Prosthetic valves

NYHA Classification

Class 1 – No breathlessness

Class 2 – Breathless on severe exertion

Class 3 – Breathless on mild exertion

Class 4 – Breathless at rest

Table 3 Modified World Health Organization classification of maternal cardiovascular risk

	mWHO I	mWHO II	mWHO II–III	mWHO III	mWHO IV
Diagnosis (if otherwise well and uncomplicated)	Small or mild – pulmonary stenosis – patent ductus arteriosus – mitral valve prolapse Successfully repaired simple lesions (atrial or ventricular septal defect, patent ductus arteriosus, anomalous pulmonary venous drainage) Atrial or ventricular ectopic beats, isolated	Unoperated atrial or ventricular septal defect Repaired tetralogy of Fallot Most arrhythmias (supraventricular arrhythmias) Turner syndrome without aortic dilatation	Mild left ventricular impairment (EF >45%) Hypertrophic cardiomyopathy Native or tissue valve disease not considered WHO I or IV (mild mitral stenosis, moderate aortic stenosis) Marfan or other HTAD syndrome without aortic dilatation Aorta <45 mm in bicuspid aortic valve pathology Repaired coarctation Atrioventricular septal defect	Moderate left ventricular impairment (EF 30–45%) Previous peripartum cardiomyopathy without any residual left ventricular impairment Mechanical valve Systemic right ventricle with good or mildly decreased ventricular function Fontan circulation. If otherwise the patient is well and the cardiac condition uncomplicated Unrepaired cyanotic heart disease Other complex heart disease Moderate mitral stenosis Severe asymptomatic aortic stenosis Moderate aortic dilatation (40–45 mm in Marfan syndrome or other HTAD; 45–50 mm in bicuspid aortic valve, Turner syndrome ASI 20–25 mm/m^2, tetralogy of Fallot <50 mm) Ventricular tachycardia	Pulmonary arterial hypertension Severe systemic ventricular dysfunction (EF <30% or NYHA class III–IV) Previous peripartum cardiomyopathy with any residual left ventricular impairment Severe mitral stenosis Severe symptomatic aortic stenosis Systemic right ventricle with moderate or severely decreased ventricular function Severe aortic dilatation (>45 mm in Marfan syndrome or other HTAD, >50 mm in bicuspid aortic valve, Turner syndrome ASI >25 mm/m^2, tetralogy of Fallot >50 mm) Vascular Ehlers–Danlos Severe (re)coarctation Fontan with any complication
Risk	No detectable increased risk of maternal mortality and no/mild increased risk in morbidity	Small increased risk of maternal mortality or moderate increase in morbidity	Intermediate increased risk of maternal mortality or moderate to severe increase in morbidity	Significantly increased risk of maternal mortality or severe morbidity	Extremely high risk of maternal mortality or severe morbidity
Maternal cardiac event rate	2.5–5%	5.7–10.5%	10–19%	19–27%	40–100%
Counselling	Yes	Yes	Yes	Yes: expert counselling required	Yes: pregnancy contraindicated: if pregnancy occurs, termination should be discussed
Care during pregnancy	Local hospital	Local hospital	Referral hospital	Expert centre for pregnancy and cardiac disease	Expert centre for pregnancy and cardiac disease
Minimal follow-up visits during pregnancy	Once or twice	Once per trimester	Bimonthly	Monthly or bimonthly	Monthly
Location of delivery	Local hospital	Local hospital	Referral hospital	Expert centre for pregnancy and cardiac disease	Expert centre for pregnancy and cardiac disease

ASI = aortic size index; EF = ejection fraction; HTAD = heritable thoracic aortic disease; mWHO = modified World Health Organization classification; NYHA = New York Heart Association; WHO = World Health Organization.

Peripartum Cardiomyopathy

Predisposing factors seem to be family history, previous history, multiparity, smoking, diabetes, hypertension, pre-eclampsia, sepsis, malnutrition, advanced age of mother and teenage pregnancy.
It is a diagnosis of exclusion when no other cause of heart failure can be found. LV may not be dilated, but LVEF is always reduced below 45%. Most common presentation is acute heart failure, but complex ventricular arrhythmias and sudden cardiac arrest are also reported.

Arrhythmias

Symptomatic palpitations can be benign but should always be investigated with ECG, ECHO and 24 Holter monitoring, especially if associated with increasing frequency or with other symptoms and signs. Not always linked to structural disease. May be related to anaemia, thyrotoxicosis, hypokalemia, or hypo/hyper magnesaemia.

- Significant arrhythmias are uncommon
- Seek expert cardiology advice for a plan
- Investigate any ongoing tachycardia; TFTs, cardiac, sepsis
- Anti-arrhythmics (major concern as adverse effects on foetus)
  - AVRT/AVNRT (Adenosine, digoxin, metoprolol, flecanide)
  - AF very rare
  - WPW (catheter ablation if poorly tolerated)
  - ICD (Presence of ICD is not a contraindication for pregnancy)
- Labour
  - Pain may be a trigger SVTs
  - Epidural with ECG monitoring
- Caesarean Section
  - Only for obstetric indications
  - Spinal/CSE safe with ECG monitoring postop

- Congenital long QT syndrome- risk of cardiac arrest is higher postpartum than during pregnancy or delivery. Beta blockers are recommended throughout pregnancy and in postpartum.
- First and second degree heart blocks often associated with CHD.

- Complete Heart block maybe congenital or acquired. May present for the first time in pregnancy, but most women with CHB will often have PPM fitted. Labour is a high risk period as the Valsalva during delivery is associated with vasovagal reaction.

Key Messages from MBBRACE –UK 2015-17

Women with Unknown Cardiac disease:

- A persistent sinus tachycardia is a 'red flag' and should always be investigated, particularly when there is associated breathlessness.

- Repeated presentation with pain and/or pain requiring opiates should be considered a 'red flag' and warrant a thorough assessment of the woman to establish the cause. Pain severe enough to prevent a woman caring for her baby represents a similar 'red flag'.

- A raised respiratory rate, chest pain, persistent tachycardia and orthopnoea are important signs and symptoms of cardiac disease which should always be fully investigated.

- It is important to be mindful of the possibility of a cardiac diagnosis when repeated attempts are made to access medical care, particularly when extreme anxiety and breathlessness are prominent symptoms.

- Syncope during exercise can suggest a cardiac origin, and should prompt cardiac evaluation.

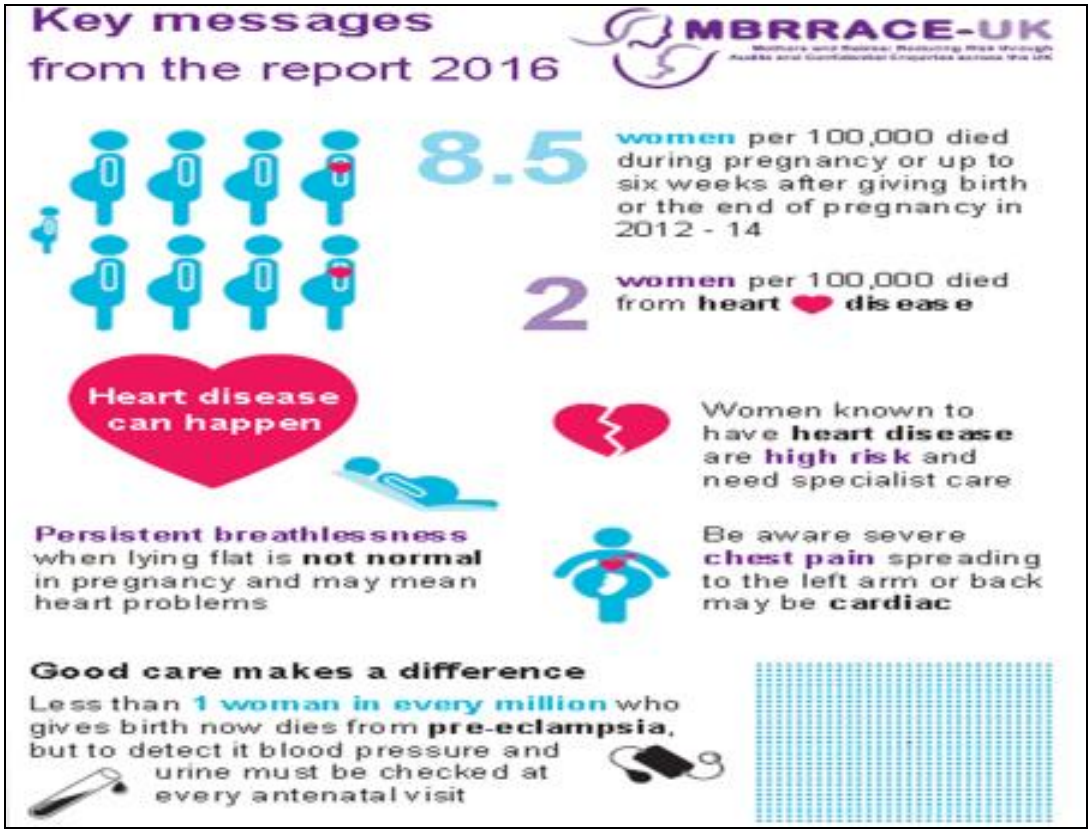
- ECG and measurement of troponin levels are recommended when a pregnant woman has chest pain. Echocardiography is recommended in any pregnant patient with unexplained or new cardiovascular signs or symptoms.

- Following resuscitation from an arrest with a likely cardiac cause, coronary angiography ± percutaneous coronary intervention is the appropriate initial diagnostic investigation.

- Electrical cardioversion is safe in all phases of pregnancy. Immediate electrical cardioversion is recommended for any tachycardia with haemodynamic instability and for pre-excited atrial fibrillation.

- When aortic dissection occurs in a young woman, the underlying diagnosis should be assumed to be an inherited aortopathy until proven otherwise.

- If there are concerns about patient compliance or access to diagnostic testing then there should be a low threshold for admission to hospital for implementation of changes to the anticoagulation regimen during pregnancy or postpartum as per ESC guidelines.



Preconceptual Counselling:

- All patients with known cardiac disease should be referred to the maternal medicine clinic for preconceptual counselling.
- Advice to be taken of current cardiac status and suitability to conceive if known to Adult Congenital Heart Disease (ACHD) services in Cardiff/ Bristol
- Arrange/ review of recent ECHO and seek advice from local cardiologist

- Offer genetic counselling if parents have Marfan Syndrome, Hypertrophic Cardiomyopathy, Long QT syndrome (autosomal dominant inheritance). Anyone with a family history or genetic confirmation of aortopathy or channelopathy should be referred for cardiac assessment before pregnancy
- Review current medication and switch to medication safe in pregnancy i.e. antihypertensives, beta blockers
- Proven SVTs should have had radiofrequency ablation of aberrant pathways
- Advise against pregnancy if has Pulmonary hypertension, LVEF<30% or NYHA 3-4, previous peripartum cardiomyopathy, severe mitral and symptomatic atrial stenosis, Marfan syndrome with aortic dilation>45, Aortic dilation >50mm with bicuspid aortic valve, native severe coarctation: discuss effective contraception
- Advise against smoking, obesity
- Achieve optimum glycemic and blood pressure control if diabetic or hypertensive
- Commence 5mg folic acid

Booking Visit:

- Women with known cardiac condition to be referred directly to Maternal medicine clinic by Community midwife (Jyoti Singh RGH, Anurag Pinto NHH)

- Ascertain cardiac condition, NYHA/ WHO classification, recent ECHO and other cardiac investigations

- Review details of recent cardiology plan if already known to cardiology

- Letter to local Cardiologist (with shortest outpatient waiting list) to inform about pregnancy and arrange review

- Referral to ACHD clinic if already known to ACHD services to enquire about cardiac status and plan place of delivery

- Book urgent ECHO/ Holter monitoring if no recent tests or new presentation (duration of Holter based on frequency of palpitations)

- Anaesthetic referral

- All **low risk** (WHO I) women are suitable to deliver in GUH after discussion with local Cardiologist

- All **moderate risk** (WHO II) women need to be discussed case by case between Obstetrician/ Cardiologist and Anaesthetist and suitability to deliver in GUH or tertiary centre to be decided on <24 weeks gestation.

- All **high risk** (WHO II-III,III,IV)women to be referred to tertiary centre for antenatal management and delivery. (Cardiac-Obstetric-Anaesthetic Clinic is held twice monthly in UHW, Wed pm for both pre-conception counselling as well as assessment/management in pregnancy).

- Termination of pregnancy can be offered to those with high risk of maternal mortality.

Antenatal Management:

- Ascertain symptoms of cardiac de-compensation: worsening palpitations, shortness of breath, chest pain
- A raised respiratory rate, chest pain, persistent tachycardia and orthopnoea are important signs and symptoms which should always be fully investigated
- Admit to antenatal ward if cardiac de-compensation suspected and request urgent cardiology review (Cardiologist of the week available in GUH 9am to 5pm)
- 60-80 % can have supraventricular arrhythmias – arrange early review with local cardiologist. Metoprolol and Bisoprolol are safe in pregnancy.
- Routine screening tests and anomaly scan, request detailed fetal cardiac examination at 22 weeks by local Radiologists for women with congenital cardiac anomalies.
- Serial fetal growth from 24 weeks
- Consider steroids if symptoms worsening and early delivery anticipated
- Ensure anaesthetic review <24 weeks gestation
- Ensure clear plan of management of labour is documented in the intrapartum record and available on CWS.
- Timing and mode of delivery should be a multidisciplinary decision depending on symptoms/ NYHA status/ Obstetric

- Vaginal delivery is well tolerated except in severe aortic stenosis, Marfan's with aortic root dilatation, aortic aneurysm, Eisenmenger syndrome, prosthetic valves on warfarin
- Any patient with new development of cardiac symptoms should have an urgent MDT review by the Obstetricians/ Cardiologists/ Anaesthetists. Cardiology investigations (ECHO/ ECG/ Holter) should be requested and decision for appropriate place and mode of delivery must be agreed on.

Intrapartum Management:

- Inform on call Obstetrician and senior Anaesthetist
- Encourage early epidural
- Ensure good blood pressure control
- Avoid dehydration and maintain input/ output chart
- Watch for and treat arrhythmias with 6mg IV adenosine
- HDU MEOWS monitoring
- Continuous EFM and maternal ECG monitoring in labour
- Restrict active second stage of labour to ≤ 60 mins (longer passive stage)

Postnatal Management:

- Avoid ergometrine and use slow IV infusion of syntocinon for 3rd stage
- Careful fluid balance
- Continue MEOWS chart and cardiac monitoring in HDU for 24 hrs
- Thrombo-embolism prophylaxis
- Good control of hypertension
- Watch for and treat arrhythmias with 6mg IV adenosine
- Neonatal cardiac check
- Discuss contraception at discharge – implanon, depoprovera
- Inform Cardiologists of delivery and arrange follow up with cardiology

References:

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