

Management of women with heart disease: in pregnancy, labour and the early postnatal period - Full Clinical Guideline

UHDB/OBS/07:23/C10

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1. Introduction

Women with cardiac disease in pregnancy are at high risk of complications and cardiac disease is the leading indirect cause of maternal mortality in the UK (MBRRACE – UK maternal report 2019)
 The Obstetric/Cardiology service at UHDB provides and offers multidisciplinary care provided by health professionals with specific interest in heart disease in pregnancy.
 Women referred should fit certain referral criteria and book under the care of a consultant with special interest or a maternal medicine consultant
 .Most patients with cardiac disease in pregnancy can expect a satisfactory outcome with careful pre pregnancy, antenatal, intrapartum and post natal management

2. Purpose & Scope

To provide guidance for care provided for women with heart disease in pregnancy.

3. Abbreviations

ABG	-	Arterial Blood Gas
ACHC	-	Adult Cardiac Health Clinic
CHD	-	Congenital Heart Disease
CXR	-	Chest X-ray
DCM	-	Dilated Cardiomyopathy
ECG	-	Electrocardiogram
ECHO	-	Echocardiogram
HDU	-	High Dependency Unit
HOCM	-	Hypertrophic Obstructive Cardiomyopathy

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IUCD	-	Intrauterine Contraceptive Device
LVEF	-	Left Ventricular Ejection Fraction
NYHA	-	New York Heart Association
PPCM	-	Peripartum Cardiomyopathy
TAPSE	-	Tricuspid Annular Plane Systolic Excursion

4. **Key Responsibilities**

This guideline is aimed for Obstetricians, Physicians and midwives providing care for pregnant women with heart disease.

5. **Preconception care**

- All women of reproductive age with congenital or acquired cardiovascular and aortic disease should have access to specialised preconception counselling in the Obstetric/Cardiac clinic
- Advice and information to empower them to make choices about pregnancy and advice about contraception and how to access services when they do become pregnant should be discussed
- Women with CHD should be seen at the joint Adult Cardiac Heart Disease Clinic (ACHD clinic) to review pre-conceptual/contraceptive issues
- All women of childbearing age seen in ACHD clinic are offered pre-pregnancy counselling appointments

6. **Termination of Pregnancy**

- Rapid access to termination of pregnancy should be facilitated, if the woman chooses this
- The pregnancy advisory clinic at Derby can be accessed for any complex cardiac condition for consideration / advice regarding Termination of pregnancy
- The termination of pregnancy service should be able to provide appropriate medical care for the severity of the woman's condition

7. **Antenatal Referral**

The outcome and safety of pregnancy is related to:

- Presence and severity of pulmonary hypertension
- Presence of cyanosis
- Functional NYHA class (New York Heart Association class), determined by the level of activity that leads to dyspnoea
- Unnecessary investigations and intervention should be avoided in Women in the low risk category
- Foetal echo, genetics referral or increased foetal monitoring may be needed
- Individual care plans should be documented in the maternity hand held records and on consultant management plan in the maternity system
- The joint obstetric and cardiology clinic occurs monthly and referrals should be directed through the obstetric consultant for triage to the clinic.
- Urgent referrals can be directed through email to the obstetric consultant for the joint clinic
- Some patients especially with complex cardiac diseases may be referred to the regional centre for assessment and / or delivery

Risk stratification:**The following patients should be counselled against pregnancy :**

Fontans procedure and additional co morbidities
 Pulmonary Arterial hypertension
 Severe systemic ventricular function (NYHA class III - IV)
 Severe coarctation
 Vascular Ehlers-Danlos
 Severe aortic dissection or history of aortic dissection
 Severe Mitral stenosis
 Patients with severe Aortic stenosis (or impaired LV function)
 Patients with previous Peripartum cardiomyopathy whose LVEF does not normalise

Predictors of maternal and neonatal events:

Predictors of maternal cardiovascular events	Predictors of neonatal events
Prior cardiac event (Heart failure/ Stroke/ Transient Ischemia / arrhythmia NYHA III/IV Left heart obstruction (moderate / severe) Reduced systemic ventricular systolic function (LVEF < 40%) Reduced sub pulmonary ventricular function (TAPSE < 16mm) Systemic Atrioventricular valve regurgitation (moderate to /severe) Pulmonary atrioventricular valve regurgitation (moderate to severe) Pulmonary Arterial hypertension Cardiac medication before pregnancy Cyanosis (O ₂ < 90%) Smoking history Repaired or unrepaired cyanotic heart disease	NYHA III/ IV / cyanosis prenatally Maternal L heart obstruction Smoking during pregnancy Low maternal oxygen saturation (< 90%) Multiple pregnancy Anticoagulation throughout pregnancy Cardiac medication before pregnancy Mechanical prosthetic valves Maternal cardiac event during pregnancy Maternal decline in cardiac output in pregnancy

LOW RISK**No detectable increased risk of maternal mortality and no/mild increase in morbidity****Relatively well and uncomplicated**

Uncomplicated small or mild : pulmonary stenosis, patent ductus arteriosus, mitral valve prolapse
 Successfully repaired simple lesions (atrial or ventricular septal defects / anomalous pulm venous drainage)
 Atrial or ventricular ectopics

MODERATE RISK

Unoperated atrial or ventricular septal defect
 Repaired tetralogy of Fallot
 Most arrhythmias
 Mild left ventricular impairment
 Hypertrophic cardiomyopathy
 Native or tissue valvular heart disease not considered WHO I or IV
 Marfan syndrome without aortic dilatation
 Aorta <45 mm in aortic disease associated with bicuspid aortic valve
 Repaired coarctation of aorta

HIGH RISK

- Mechanical valve
- Systemic right ventricular hypertrophy
- Fontan circulation
- Cyanotic heart disease (unrepaired)
- Other complex congenital heart disease
- Aortic dilatation ≥ 40 mm in Marfan syndrome
- Aortic dilatation ≥ 45 mm in aortic disease associated with bicuspid aortic valve

**EXTREMELY HIGH RISK of maternal mortality or severe morbidity.
ADVICE AGAINST PREGNANCY**

- Pulmonary arterial hypertension of any cause
- Severe systemic ventricular dysfunction (LVEF $< 30\%$, NYHA III–IV)
- Previous peripartum cardiomyopathy with any residual impairment of left ventricular function
- Severe mitral stenosis, severe symptomatic aortic stenosis
- Marfan syndrome with aorta dilated > 45 mm aortic dilatation > 50 mm in aortic disease associated with bicuspid aortic valve
- Native severe coarctation

8. Antenatal care

- Early consultant booking for management plan at 12 week
- Women with CHD should have a consultant cardiac scan at 20-22 weeks
- Cardiovascular assessment of pulse rate/rhythm, auscultation for change in murmur and lung changes associated with pulmonary oedema should be done at each visit
- Women with cyanotic heart disease should have their oxygen saturations checked at each trimester or more in any clinical signs of deterioration
- Echocardiography should be performed in women WITH unexplained or new cardiovascular signs or symptoms
- All patients with cardiac conditions should be offered an anaesthetic opinion and patients in the moderate to high risk group above should have an anaesthetic clinic referral and a clinic appointment

General principles of management:**Dysrhythmias**

- Sinus tachycardia and palpitations may be a normal feature of pregnancy.
- Ectopic beats are common in both mother and fetus and generally have no adverse effects on either.
- Rule out anaemia, thyroid disease, together with an examination of cardiovascular and respiratory symptoms.
- Referral to the Joint cardiology clinic should be for those with ECG abnormalities, severe dysrhythmias or who are symptomatic (e.g. syncope or severe breathlessness).

Coronary Artery Disease

The diagnostic criteria for Coronary Heart Disease in pregnancy and puerperium is similar to that in non-pregnant population

- chest pain
- ECG changes
- Cardiac biomarkers (troponin 1)

Consider the diagnosis especially if related to other risk factors like smoking, hypertension, obesity, hyperlipidaemia, older age, diabetes mellitus, and a positive family history

Differential diagnoses include pre-eclampsia, pulmonary embolism, and aortic dissection

Raised / increased Troponin levels should instigate referral to cardiologist / interventional cardiology for review

- ECHO to check for wall motion abnormalities may be useful in addition to other investigations

Cyanotic congenital heart disease

- Significant risk for mother and fetus
- May risk worsening cyanosis, VTE, Fetal growth restriction (FGR), IUFD
- Associated pulmonary hypertension

Cardiomyopathy and heart failure

- **Hypertrophic obstructive cardiomyopathy (HOCM)**
 - Mostly well tolerated in pregnancy
 - May require B blockers in pregnancy for symptom control
 - Avoid hypotension
- **Dilated cardiomyopathy (DCM)**
 - Condition can worsen in pregnancy and postpartum. This can be irreversible
 - Careful monitoring of symptoms
 - ECHO to monitor function
- **Peripartum cardiomyopathy (PPCM)**
 - Presents with heart failure and Left ventricular systolic dysfunction in late pregnancy and in the months following delivery
 - Treat in conjunction with Cardiology and anaesthetic support
 - Symptoms of heart failure (similar to any presentation including DCM)
 - Dyspnoea
 - Pulmonary odema
 - Peripheral odema
 - Palpitations
 - Tachycardia
 - Tachypnoea
 - Cardiac failure
 - Dysrhythmias
 - ECHO for diagnosis
 - Treat for heart failure
 - Thromboprophylaxis

Thoracic Aortic dissection

- **Haemodynamic and vascular changes increase risk in certain conditions**
 - Marfans syndrome
 - Turners syndrome
 - Aortic dilatation with bicuspid Aortic valve
 - Aortic dilatation with coarctation of Aorta
 - Vascular Ehlers – Danlos syndrome (type IV)
- Severe Acute chest pain radiating to the scapulae and systolic hypertension
- Regular ECHO throughout pregnancy to assess
- May require Beta blockers
- Caesarean delivery if significant aortic root dilatation (> 4cm)

Previously undiagnosed heart disease

We all should be aware that in anyone with the following symptoms and/or signs, cardiac disease should be considered

- Unexplained tachycardia
- Hypotension
- New onset shortness of breath
- Chest pain
- Palpitations associated with symptoms such as collapse or syncope

Such patients **may** require ECG, Echo, CXR, ABG and cardiology review as part of their investigation

9. Labour

- Patients who require any specific management in labour will have a care plan filled out (see appendix A)
- If a woman tolerates pregnancy well then she is likely to tolerate delivery
- On admission to the labour ward refer to plan of care and inform
 - Senior obstetrician
 - Senior anaesthetist
- **Vaginal delivery** is recommended as **first choice** in most women
- Epidural anaesthesia and assisted vaginal delivery is appropriate where necessary
- Caesarean section is usually necessary **only** for obstetric indications
- Avoid fluid overload
- Fluid balance should be adjusted for the particular disorder.
- Central venous lines and an arterial line at anaesthetic discretion
- Maternal monitoring - Depending on nature of the cardiac disease and decided by the anaesthetic staff.
- Continuous electronic foetal monitoring is recommended
- Involve the anaesthetic staff early in the management of labour
- Appropriate analgesia is important to prevent problems caused by increased cardiac output in labour
- Some women will require antibiotics at delivery and this is based on the risk of endocarditis
 - **Antibiotic prophylaxis is not recommended routinely for the prevention of endocarditis**
 - However any infection in a patient at risk of endocarditis should be investigated promptly and treated appropriately with consultation with the consultant microbiologist

For some women, a short active second stage or no active second stage may be necessary and vaginal delivery should be assisted. If needed this will be specified in the care plan

- Active management of the 3rd stage
- Syntocinon 5units in 20ml saline over 20 min to be used
- For Post-partum Haemorrhage due to uterine hypotonia
 - Use mechanical methods
 - e.g. bimanual compression
 - Brace suture
 - Use misoprostol in preference to haemobate

10. Post Natal Care

Extended post natal care may be required in HDU

Monitor fluid balance

Women should be supported with breast feeding

11. Contraception

- Prior to discharge contraceptive options should be discussed

12. Monitoring Compliance and Effectiveness

As per Business Unit audit forward programme

13. References

- RCOG (2011). Good Practice No.13, June 2011, Cardiac Disease and Pregnancy, RCOG
- NICE (2008). Prophylaxis against infective endocarditis: NICE Guideline ,March 2008
- MBRRACE-UK: Saving Lives, Improving Mothers' Care December 2019
- ESC Guidelines on the management of cardiovascular diseases during pregnancy 2018

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Clinical management / plan for delivery for moderate to high risk women

Cardiac diagnosis.....
 Place of proposed delivery: Burton Hospital / Derby Hospital

If admitted to LW, please inform	Consultant obstetrician on call	Yes / No
	Obstetric SpR on call	Yes / No
	Consultant anaesthetist on call	Yes / No
	Anaesthetic SpR on call	Yes / No
	Cardiologist informed	Yes / No
Mode of delivery		
Elective LSCS	Syntocinon reduced regime (See Pre-eclampsia guideline)	Yes/no
	Anaesthetic technique	Epidural/Spinal /GA
Vaginal delivery: 1st stage	TED stockings in labour	Yes / No
	HDU chart	Yes / No
	Maternal monitoring – ECG / SaO2 / arterial line BP / CVP Continuous EFM is recommended for all women with moderate to high risk	Yes / No
Vaginal delivery : second stage	Normal second stage	Yes / No
	Elective assisted delivery only	Yes / No
	Consider assisted delivery if not delivered in 60 minutes	Yes / No
Vaginal delivery : 3rd stage	Active Management	Yes / No
	Syntocinon infusion – 5 units in 20 mls saline over 20 min DO NOT GIVE ERGOMETRINE	Yes / No
Postpartum	High-dependency unit	Yes / No For hours
	Thromboprophylaxis	Yes / No
	Medications to be given	Name
	Continued for	Dose.....Duration.....days/weeks
	Cardiac review	Yes / No at weeks.....
	Contraceptive plans discussed	Yes / No

Postpartum haemorrhage	Syntocinon - reduced regime	Yes
	Misoprostol 600 micrograms PR	Yes
	Consider use of intrauterine balloon (antibiotic cover is recommended)	Yes
	Input/output charts to be maintained	Yes
	Consider central access or arterial monitoring Avoid Hemabate or high-dose Syntocinon	Yes
Preterm labour	Atosiban (Tractocile)	Yes
Pacemaker	Do not use unipolar diathermy Beware pacemaker in unusual places (e.g. abdominal wall) at LSCS	De-activate implantable defibrillators

Please inform the consultant obstetrician on call if there is departure from planned management or if unexpected clinical situations develop in women with cardiac disease.

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Contraception in women with heart disease

If you know you have a heart problem, it is important that you have the opportunity to discuss fertility, pregnancy and contraception with a specialist who knows how your condition affects the safety of the various options available so they can advise you about the methods most suitable to you. By planning ahead you will avoid having to deal with the crisis of an unexpected pregnancy.

The first question to answer when considering what contraceptive to use is: **what are the risks for me if I become pregnant?** Some women will be very high risk and therefore will need contraception that is very effective at preventing an accidental pregnancy. Women at lower risk may be willing to accept a contraceptive method with a higher failure rate.

The perfect contraceptive has not been invented – all have advantages and disadvantages. No contraceptive is 100% reliable (even sterilisation). This leaflet outlines some of the options available. However, to be sure that you choose the right method, it is vital that you discuss your individual case with a heart/pregnancy specialist.

Natural methods

There are a variety of techniques that use our understanding of what time in the cycle conception occurs to try and prevent pregnancy. These methods are not very reliable and depend very much on how carefully they are used. They don't have any adverse effects, but if it is really important that you don't get pregnant, these methods are not for you.

Barrier methods (condoms, diaphragm)

Like natural methods, barrier contraception has few adverse effects but again has a high failure (pregnancy) rate even when used with spermicidal creams. However, condoms have the additional benefit of protecting against sexually transmitted diseases.

Coils (intrauterine contraceptive devices, IUCDs)

Coils, or IUCDs, are much more reliable, with as few as one woman in 100 getting pregnant over a period of five years. There are two types: those wrapped in copper (e.g. TT380) and those containing a hormone similar to progesterone (e.g. Mirena). The Mirena coil has the advantage of causing less bleeding (periods often stop entirely) and less infection than copper coils, and can therefore be used more safely in women who have never had children (whose wombs are more at risk of infection).

About one in 1000 women have a fainting reaction at the time the coil is inserted. This can be dangerous for women with severe heart disease if there is no expert help available. So, if a coil is to be used, it should be inserted in hospital, with cardiac anaesthetic expertise on standby in case of this rare complication (an actual anaesthetic is not usually necessary). A rare complication of all coils is pregnancy in the fallopian tube (ectopic pregnancy), which usually have to be removed surgically. However, the risk of pregnancy is extremely low with the Mirena coil (even lower than after sterilisation). Mirena coils are effective for up to five years.

Oral contraceptive pills

There are two main types of oral contraceptive pills: those with both estrogen and progestogen hormones (the combined pill) and those with only a low dose of progestogen (the low-dose or mini pill).

The **combined pill** is probably the most effective, with failure rates of less than one in 300 women per year if taken correctly. It has many advantages, especially regulating the periods. The combined pill is also available as a patch or vaginal ring. Good Practice No. 13 6 of 18 © Royal College of Obstetricians and Gynaecologists

The most important complication of the combined pill is that it can cause blood clotting or thrombosis (three- to four-fold increased risk). Thrombosis happens in about one in 5000 women per year. One-quarter of these clots will be fatal. This risk (for the average woman) is still only about half that of

dying from being pregnant. Certain heart conditions are associated with an increased risk of clotting and therefore you may be told that this form of contraception is not suitable for you.

By contrast, **the traditional low-dose or progestogen-only pill** has almost no dangerous adverse effects and does not cause thrombosis. However, it has a higher failure rate than the combined pill.

As a result, a newer progestogen-only pill containing desogestrel which stops ovulation (Cerazette) is used. There is also a longer window of time for the woman to remember to take her pill, so the occasional missed pill is less likely to result in pregnancy. Cerazette is related to the drug in Implanon® and can be used as a test before the implant is inserted. About one in five women discontinue Cerazette because of irregular bleeding.

Progestogen-only injectable (depot) injections of hormone (Depo-Provera®) These are intramuscular injections of progestogen which last for 12 weeks. Periods will often disappear, although they may be irregular or heavy for a while when you decide to stop the injections. The failure rate is low: only one pregnancy in 300 women per year. Injections might be a problem if you are on warfarin or have a needle phobia.

Implant of progestogen (or Nexplanon®)

This is a small implant which is inserted under the skin in the upper arm by a doctor or nurse. It delivers a continuous dose of the hormone etonogestrel. In one in five women, periods stop completely. Implanon is one of the safest and most effective forms of contraception available. The implant is effective for three years. The risk of getting pregnant is very low: less than one in 1000 per year. Nexplanon has replaced Implanon, which was sometimes difficult to insert correctly.

Caution: The drug bosentan, sometimes used for heart disease, can reduce the effectiveness of most hormonal contraception, including Cerazette and Nexplanon, so additional contraception should be used if you need to take bosentan.

Sterilisation

Some couples decide that they don't want to become pregnant at any point. If so, sterilisation is an option. Both men (vasectomy) and women can be sterilised. Vasectomy is more reliable and safer. In women most sterilisations are performed with clips applied to the tubes. This can be done by a keyhole method (laparoscopy) under anaesthetic. A mini-laparotomy (proper scar rather than a keyhole incision) under a regional anaesthetic (not asleep) may be safer for some women with heart problems (laparoscopy involves putting gas at high pressure into the abdomen so that the womb and tubes can be visualised, and this can affect the heart).

The risk of getting pregnant once the clips have been applied is only about one in 500 (pregnancy can occur if the clip does not close the tube). The tubes can be cut and tied at caesarean section, but then the risk of the tubes joining up again is greater, about one in 200.

What about emergency contraception, including the 'morning after pill'?

Emergency contraception can be used up to five days after unprotected sex, a burst condom or missed pills. The copper IUCD (coil) is the most effective method of emergency contraception and will prevent over 99% of pregnancies. It can sometimes be used later than five days after sex, if it is likely to be no more than five days since you released an egg (ovulated). You may be offered an antibiotic to prevent pelvic infection. You can choose to keep using the IUCD for contraception, or it can be easily removed when your next period comes.

Oral emergency contraception (the 'morning after pill') can be used up to five days after sex. The sooner it is taken, the more effective it is likely to be. There are two types of pill available.

One contains progestogen hormone (levonorgestrel) and is available to buy or sometimes free of charge from pharmacies (Levonelle®). Levonelle can be used up to 72 hours after sex. It may upset warfarin control. It is not advisable if you have a rare condition called porphyria (nothing to do with heart disease). It is one tablet which you take as soon as possible.

The other pill is a drug called ulipristal acetate (ellaOne®), which can be used up to five days (120 hours) after sex and is available on prescription from your local doctor or sexual health clinic. It should not be used by women with severe asthma or liver disease.

The adverse effects of emergency oral contraceptive pills are mild (nausea, breast tenderness, disruption to periods) and there are no long-term effects. The efficacy of Levonelle and ellaOne can be affected by some medications.

Other sources of information

- Family planning clinics and family doctors
- Grown Up Congenital Heart Patients Association: www.guch.org.uk (congenital heart disease)
- FPA: www.fpa.org.uk
- New York Online Access to Health: www.noah-health.org/en/pregnancy/contraception (general)
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	1	1998	Maternity Development Committee	New
	2	2000	Miss Fowlie (Consultant) Dr McCance (Cardiologist)	Review
	3	2009	Miss Hamilton (Consultant) Dr Salah (Registrar) Dr McCance (Cardiologist)	Update / Publication of NICE guidance
	4	2013	Miss Rajendran Dr Hazra O&G StR Dr A McCance Cardiologist	Review and Updated based on RCOG Good Practice & European society of Cardiology Guidelines 2011
UHDB	1	April 2020	Miss S Rajendran – Consultant Obstetrician	Review / Update
	1.1	Aug 2021	Miss S Rajendran – Consultant Obstetrician Dr A McCance Cardiologist	ACHD paragraph 5 clarified pre-pregnancy counselling
	2	Mar 2023	Miss S Rajendran – Consultant Obstetrician Cardiologist	Review
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