Royal College of
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Cardiac Disease and Pregnancy

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1. Purpose

Cardiac disease is a leading cause of maternal death in pregnancy in many developed countries, including the UK. However, there is a lack of evidence-based guidelines to assist in planning the management of affected pregnancies. The purpose of this Good Practice guidance is to provide a summary of current expert opinion as an interim measure, with the hope that these opinions will be supplemented by objective evidence in due course.

2. Introduction

In the UK, the Confidential Enquiries into Maternal Deaths (CEMACH) have shown that the overall rate of mortality from cardiac disease has risen from 7.3/million births in the 1982–84 triennium[1] to 22.7/million births in the 2003–05 triennium.[2] The major part of this increase is attributable to acquired heart disease, deaths from which have risen from 4.7/million births to 20.8/million births. One-third of these deaths are a result of myocardial infarction/ischaemic heart disease and a similar number of late deaths are associated with peripartum cardiomyopathy. Other significant contributors (5–10% each) are rheumatic heart disease, congenital heart disease and pulmonary hypertension. With the current increase in older mothers, obesity, immigration and survival of babies operated on for congenital heart disease, the need to identify women at risk of heart disease and to plan their careful management will also inevitably increase.

The suggestions in this Good Practice guidance are based upon the recommendations of a consensus group convened at the Royal College of Obstetricians and Gynaecologists in 2006, which are published in full by the RCOG Press,[3] and those in the CEMACH report *Saving Mothers' Lives: Reviewing Maternal Deaths to Make Motherhood Safer 2003–2005*.[2]

3. Risk assessment, preconception counselling and management planningn

3.1 Myocardial infarction, ischaemic heart disease, and aortic dissection

Pregnancy itself raises the risk of acute myocardial infarction by three- to four-fold, with the risk being 30 times higher for women over the age of 40 years compared with women aged less than 20 years. As reported by CEMACH, in the 2003–05 triennium[2] the rate of maternal death from ischaemic heart disease in the UK had risen to 1/132,000 pregnancies (up from 1/252,000 in 2000–2002).[4] Other risk factors include chronic hypertension, pre-eclampsia, diabetes, smoking, obesity and hyperlipidaemia. Up to 1/13 women with a myocardial infarction in pregnancy will die. Unfortunately, many of these risk factors are becoming increasingly common, and most women affected will be asymptomatic before pregnancy, with no history of heart disease. The key component of good management is therefore a high index of suspicion for myocardial infarction in any pregnant woman presenting with chest pain. The possibility of aortic dissection should also be considered. All women with chest pain in pregnancy should have an electrocardiogram interpreted by someone who is skilled at detecting signs of cardiac ischaemia and infarction and, if the pain is severe, they should have computerd tomography or a magnetic resonance imaging scan of the chest. A serum troponin I measurement can also be useful.

3.2 Peripartum cardiomyopathy

The cause of peripartum cardiomyopathy is unknown in most cases. It usually presents in late pregnancy or early in the puerperium, but it can occur up to 6 months after delivery. Peripartum cardiomyopathy should be considered in any pregnant or puerperal woman who complains of increasing shortness of breath, especially on lying flat or at night. As 25% of affected women will be hypertensive, it can be confused with pre-eclampsia. All such women should have an electrocardiogram, a chest X-ray and an echocardiogram.

3.3 Rheumatic heart disease

Almost 25% women currently giving birth in the UK were not themselves born in the UK; the figure rises to more than 50% in London. Many of these women will never have undergone medical screening and some will be unaware that they have valvular heart disease. This highlights the need for a particularly careful cardiovascular assessment at the beginning of pregnancy of all women not born in a country where there is effective medical screening in childhood, including auscultation of the heart. Mitral valve stenosis (the most common lesion and the one that carries the highest risk) is a difficult clinical diagnosis and there should be a low threshold for echocardiography.

3.4 Aortic dissection

Systolic hypertension was a key factor in most of the deaths from aortic dissection, and this emphasises the importance of blood pressure monitoring during pregnancy and prompt antihypertensive therapy if blood pressure becomes elevated. Aortic dissection (diagnosed by computed tomography scan) is the most common serious complication of Marfan syndrome.

3.5 Congenital heart disease

Although deaths from congenital heart disease are fortunately uncommon, the prevalence of this condition in pregnancy is about 0.8%. Congenital heart disease is one of the most common congenital abnormalities and the majority of those affected will survive to adulthood, in large part because of the development of effective corrective/palliative surgery over the last 30 years.

3.6 General principles of management

3.6.1 Preconception

Girls with congenital heart disease should be referred to a joint cardiac/obstetric/gynaecological clinic for advice about contraception (and subsequently for preconception counselling) once puberty is under way (commonly at the age of 12–15 years). Preconception counselling should also be offered to older women with a new diagnosis. Because pregnancy carries substantially increased risks for women with congenital heart disease, particular efforts should be made to prevent unwanted pregnancy. Appendix A describes appropriate types of contraception for women with the different types of congenital lesion.

Women should be given an outline of the issues relating to pregnancy with congenital heart disease at the first visit to the joint clinic, and then be reviewed with more detailed information once they are considering conception. Topics that should be covered at this detailed review include the increased risk of mortality, congenital heart disease in the offspring and the need for increased medical surveillance during pregnancy. A sample patient information leaflet on congenital heart disease and pregnancy is available in Appendix B. Appendix C describes what to look out for in the symptomatic women. Appendix D describes the typical patient journey of a pregnant woman with heart disease.

3.6.2 Antepartum

Because there are so many types of cardiac disease, often with very different implications, it is important that a risk assessment of any woman with a heart murmur or a history of any cardiac defect should be carried out early in pregnancy in a joint clinic attended by a consultant obstetrician, cardiologist and anaesthetist. All of these professionals should have appropriate experience. Women at low risk can be identified and returned to routine care. Women at significant risk of adverse events during pregnancy should be seen regularly in the antenatal clinic, whenever possible by the same consultant obstetrician, who should have appropriate competencies in this field. When that consultant is away on leave, there should be a competent deputy. Use of the structured antenatal notes makes continuity of care easier to achieve. Cardiovascular assessment should be carried out at every antenatal clinic. Blood pressure should be measured manually with a sphygmomanometer according to the recommendations of the British Hypertension Society (http://www.abdn.ac.uk/medical/bhs/). Measurement of pulse rate and rhythm is also mandatory as it may

be the first sign of volume overload. Auscultation to assess any change in murmur or any lung changes associated with pulmonary oedema is recommended in all cases of significant cardiac compromise (which will have been identified early in pregnancy at the joint clinic). Women with cyanotic heart disease should have their oxygen saturations checked periodically (each trimester or more often if there are any clinical signs of deterioration). A template for adapting normal antenatal records for use in women with heart disease is available in Appendix E.

All women with structural congenital heart disease should be offered a fetal echocardiogram during the second trimester to be carried out by an accredited paediatric/fetal cardiologist (as distinct from the standard four-chamber view offered to all women as part of routine antenatal screening and carried out by accredited ultrasonographers and fetal medicine specialists).

A further multidisciplinary meeting should take place at 32–34 weeks of gestation to establish a plan of management for delivery. Important features of such a plan include deciding who should be involved in supervising the labour, whether a caesarean section is appropriate, whether bearing down is advisable in the second stage and appropriate prophylaxis against postpartum haemorrhage (routinely used oxytocic regimes can have major cardiovascular adverse effects; a low-dose syntocinon infusion is probably the safest option, and at caesarean section prophylactic uterine compression sutures can be considered instead of oxytocics). The plan should also include postpartum management, including whether prophylaxis against thrombosis is appropriate, the length of postpartum stay in hospital and the timing of cardiac and obstetric review. A template for such planning is provided in Appendix F.

3.6.3 Intrapartum

The general principle of intrapartum management is to minimise cardiovascular stress. In most cases this will be achieved by the use of early slow incremental epidural anaesthesia and assisted vaginal delivery. Caesarean section is usually necessary only for obstetric indications.

Some women will benefit from specialist care at tertiary units. The decision about the optimum place for antenatal and intrapartum care should be made in conjunction with obstetricians and cardiologists at tertiary units known to specialise in the management of women with heart disease in pregnancy. Appropriate tertiary units will have high-dependency and intensive care units suitable for the care of pregnant women with significant heart disease.

3.6.4 Postpartum

The length of recommended stay in hospital and any suggested special measures (such as anticoagulation, or observation in a high-dependency area) should be specified in advance, so that midwifery/resident medical staff do not have to seek urgent guidance out of hours. The timing of follow-up at the joint clinic should also be specified. Appropriate advice about contraception should also be given.

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The final version is the responsibility of the Safety and Quality Committee.

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The RCOG will maintain a watching brief on the need to review this guidance.

APPENDIX A

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 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.

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 woman 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 (e.g. Micronor) 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, Micronor is being replaced by a new version, Cerazette® (ORGANON Labs Ltd), containing desogestrel, a newer progestogen-only pill which stops ovulation. 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.

A technique that has recently become available involves putting tiny implants into the fallopian tubes to block them. This is done via a hysteroscope (a small telescopic microscope which is passed through the vagina and cervix to look inside the womb). This can be done under local anaesthetic or intravenous sedation, although it should always be done in a centre fully equipped to deal with women with heart problems. It is called Essure. Essure is not yet widely available, so your doctor should advise you where it can be done.

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). You can buy this pill from the pharmacist without a prescription (cost in 2009: £22); 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)

My heart condition: _____

Contraceptive priorities: _____

Specific risks: _____

Recommendations: _____

In general, the most effective contraceptives should be used in women with the most serious heart disease as the consequences of failure (pregnancy) are far greater.

The RCOG is grateful to Dr Lorna Swan (Royal Brompton Hospital) and Professor Phil Steer (Chelsea and Westminster Hospital) for permitting the adaptation of their local leaflet.

APPENDIX B

Patient information on congenital heart disease and pregnancy

About eight in every 1000 babies born has a congenital heart defect, and three of these will be severe. Many can be helped by surgery, which has improved enormously over the last 50 years. Eventually they will grow up and many will want to have children of their own. If you have grown-up congenital heart disease (GUCH) and want to be (or are) pregnant, you should consider the following:

See your cardiologist

The cardiologist is the expert in heart disease, and their job is to keep you as well as possible. They will know the details of your condition, and they can explain to you the effect that pregnancy might be expected to have on your health. If they think pregnancy will be dangerous for you, they may advise you not to become pregnant. However, you should remember that ultimately this is a decision only you can make, in conjunction with your partner and in the knowledge of all the facts.

It is very important that full testing is carried out before pregnancy to establish how well your heart is working. This will enable the cardiologist to give you the most accurate advice, and the information gained will be vital in the proper care of a pregnancy. Some tests, such as X-rays and cardiac catheterisation, are best avoided in pregnancy and so if necessary should be done before conception. Pregnancy puts quite a strain on the heart, and sometimes surgery to improve its function can be undertaken which will make a subsequent pregnancy safer.

See the obstetrician before you become pregnant

The obstetrician is the expert in pregnancy. It is important that you see one with an understanding of the special needs of GUCH women who are pregnant. This may mean attending the specialist centre in your region, which is likely to be a teaching hospital. Your cardiologist should know of an obstetrician with the relevant experience and skills. The obstetrician will need as much information as possible about your heart, so it is a good idea to get a full report from your cardiologist to take with you and, if possible, the report of a recent echocardiogram. Ideally, you should see the obstetrician and cardiologist together, at a joint clinic.

The obstetrician's job is to be supportive of women who want to be or are pregnant. The cardiologist might say that you have a 5% risk of not surviving a pregnancy; the obstetrician is more likely to say you have a 95% chance of surviving. You will need to balance carefully what both the cardiologist and obstetrician say and be aware of their different points of view. You need to be aware that all women who become pregnant face a small risk of dying as a result of their pregnancy – in the UK it is currently about one chance in 8000. However, in some cases of very severe heart disease (such as Eisenmenger's syndrome or primary pulmonary hypertension), the risk of death is as high as 25–40% (one in four to almost one in two). It is often difficult to give a precise estimate of risk for the more unusual forms of heart defect. In addition, the chance of dying is exactly that – a chance. Some women with a very high risk will survive, and some with a very low risk will die. You need to discuss with your partner and your family what risk you are prepared to take.

The heart pumps blood around the body, and the blood carries oxygen and nourishment. If the pump does not work as well as normal, the developing baby may not get all the oxygen and food it needs. It may therefore not grow as well as normal (fetal growth restriction) or it may be born premature (or 'preterm' as we now say). With good neonatal care, many small babies can do well after they are born, but some may have a permanent handicap. For a few, this will be severe. You need to consider how you would cope with this if it happens.

In addition, the tendency to have a heart defect is hereditary; if you have one your baby will probably have a 3–5% risk (one in 20) of having one too (the risk varies somewhat, depending on the precise condition). This is about five times the average risk. If your partner has a heart problem, the risk is even higher. Nowadays, up to 80% of heart abnormalities can be detected using ultrasound scanning. This will be offered to you between

11 and 24 weeks of the pregnancy (the later the scan, the bigger the baby, and the more detail can be seen). If an abnormality is detected, you will be offered the possibility of terminating the pregnancy. You will need to decide how you feel about this.

These days, much medical care, including antenatal care, is done as an 'outpatient'. However, if your heart has difficulty pumping well enough to meet both your needs and the needs of the developing baby, extra rest will be necessary. Sometimes, adequate rest can be obtained only by admitting the mother to hospital, where she needs to do nothing except grow the baby. In addition, close observation of your heart and of the developing baby may be necessary on a day-to-day basis. All this means that you need to plan for the possibility of spending quite a lot of time in hospital, and in a few cases this can be most of the pregnancy.

Sadly, some forms of GUCH mean that life expectancy is reduced. You should think about what will happen to your child if you die early. A supportive family structure is very helpful in safeguarding the child's interests.

What will happen when I am pregnant?

The demand on the heart increases from very early pregnancy, as the hormones adjust the mother's body to help the developing baby (fetus). You should see your obstetrician very early (at about eight weeks from the beginning of the last period, which is about six weeks from conception of the baby). Your pregnancy should be jointly supervised by a cardiologist and an obstetrician, ideally at the same clinic. It is very important to see the obstetrician frequently, so that they can get used to you and how you are, and you can get to know them. This way, they will be much more able to pick up early signs of any problem developing. Depending on her cardiac status, the woman should be seen by an appropriately experienced consultant obstetrician every two to four weeks until 20 weeks, then every two weeks until 24 weeks, and then weekly thereafter.

At each visit, you will be asked about shortness of breath (especially at night) and your exercise tolerance (can you still climb stairs or walk at your normal pace), palpitations (irregular heart beat) and your own feelings of how things are going (for example, are you feeling the baby move). They will measure your pulse rate and rhythm, your blood pressure, whether you have any fluid collection at the ankles (oedema) and the size of the uterus to judge how well the baby is growing. They will also listen to your lungs (again to check for any collection of fluid, or pulmonary oedema) and your heart (to detect any changes in murmurs which might indicate a deterioration in the functioning of a valve, or infection of the heart). You will also see a midwife who will advise you about the normal aspects of pregnancy and birth.

It is important to minimise the strain on the heart by vigorous treatment of any infections (for example chest, urinary). If the heart beat has any tendency to be irregular, drugs such as atenolol or digoxin may be given to control the rate. Regular scans to check on the growth of the baby will probably be necessary. If there is any anxiety about your condition, or that of your baby, you are likely to be admitted to hospital for rest and tests.

What about the birth?

Natural labour, if it goes well, is the safest way for anyone to give birth, and this includes women with GUCH. The main aim is to limit the demands on the heart, and for this reason good pain relief (usually with an epidural, an injection of local anaesthetic around the spinal cord) is very important. Also, bearing down ('pushing the baby out') at the end of labour can be very exhausting, so it is often recommended that this part is assisted by the doctors (using either a suction cup or forceps on the baby's head). Caesarean section may be advised for the same reasons as in other pregnant women. Antibiotics are occasionally given to prevent infection of the heart (although they are not necessary if the birth is entirely normal, whereas they are routine anyway if delivery is by caesarean section).

When can I go home?

Women with GUCH usually need to stay in for a few days longer than average, to allow their heart to adjust to them not being pregnant. Clots (thrombosis) in the veins of the leg are more common after birth, especially in women with GUCH, and you will probably be given injections to thin the blood slightly (heparin) until

you are fully mobile. It is important to consider starting contraception before your fertility returns. This may be as early as four weeks after delivery if you are not fully breastfeeding.

And finally...

Don't forget that if you decide to get pregnant, taking extra folic acid (easily obtainable from most pharmacies) for three months before and after conceiving will reduce substantially the risk of the baby having spina bifida (this applies to all women, not just those with heart disease). You should also make sure you have a good diet, and aim for a good body weight (not too fat or too thin). It is also advisable to get a blood test from your doctor to make sure that you are immune to rubella (German measles), because if you are not, it is a good idea to be vaccinated before you become pregnant (rubella is very dangerous to the baby if you become pregnant). And of course, if you are a smoker, you should do your very best to stop before you become pregnant.

The RCOG is grateful to Professor P Steer FRCOG (Imperial College London, Chelsea and Westminster Hospital) for permission to use this patient information leaflet.

APPENDIX C

Pregnancy in women with heart disease: what to look out for in the symptomatic woman

Many women with cardiac disease will be symptomatic before they become pregnant. It is important that everyone caring for the woman during pregnancy is aware of her prepregnancy symptoms, firstly so that they do not overreact to similar symptoms during pregnancy, and secondly so that they can detect as soon as possible any deterioration in symptomatic status. The New York Heart Association (NYHA) classification is as follows:

NYHA class	Symptoms
I	No symptoms and no limitation in ordinary physical activity, e.g. shortness of breath when walking, climbing stairs, etc.
II	Mild symptoms (mild shortness of breath and/or angina) and slight limitation during ordinary activity.
III	Marked limitation in activity due to symptoms, even during less-than-ordinary activity, e.g. walking short distances (20–100 m). Comfortable only at rest.
IV	Severe limitations. Experiences symptoms even while at rest. Mostly bedbound patients.

Many pregnant women will experience deterioration of one class as pregnancy progresses, and they should be warned about this. They may need to take more rest than usual during pregnancy, although it is also important for them to maintain their fitness as much as possible. Clinicians should be familiar with the appropriate questions to elicit symptoms accurately. For example, in response to the question 'do you get short of breath climbing stairs?', the answer may be 'no', because the woman has stopped climbing stairs. The correct question is 'how many flights of stairs can you climb at a steady pace without having to stop because of shortness of breath?'.

Most pregnant women complain of tiredness, and women with cardiac disease are no exception. This is why continuity of carer is so important, because sometimes deterioration in the woman's condition is more apparent in her demeanour and the way she answers questions than in the precise answers she gives. A useful tactic is to call a woman to your consulting room yourself and watch how quickly she can walk from the waiting area to your consulting room, how short of breath this makes her, and what her pulse rate and rhythm is when she first sits down (a 'mini exercise test').

A rising pulse rate can be one of the first signs of cardiac decompensation. The pulse rate is best measured using a stethoscope and auscultating the heart, because when the pulse becomes fast, irregular or faint, the radial pulse is often difficult to detect accurately.

The woman's blood pressure should be checked carefully using a manual sphygmomanometer. The woman should be seated comfortably, not talking, with an appropriately sized cuff placed on the correct arm (for example, the right arm is usually used in women with coarctation of the aorta, 80% of whom will also have a bicuspid aortic valve). The arm should be supported and held out at an angle so that the cuff is at the level of the left atrium. An excellent resource showing how the blood pressure should be taken correctly can be found at http://www.abdn.ac.uk/medical/bhs/booklet/proced.htm

The heart sounds should be auscultated carefully at each visit in a standard place, commonly the left sternal edge, to check if there has been any substantial change from the previous visit. Heart murmurs are graded from one (extremely soft) to six (the loudest one has ever heard). It is usual for a murmur to increase by one grade as pregnancy progresses because of the increase in cardiac output. A sudden increase in the loudness of a heart murmur can suggest the development of vegetations from endocarditis. The appearance of a new murmur is nearly always significant. For example, in a woman with Marfan syndrome, the appearance of a diastolic murmur can indicate dilatation of the aortic root with the onset of aortic regurgitation. This will usually require urgent intervention as it may lead to heart failure or aortic dissection.

The lung bases posteriorly should be auscultated at each visit to check for crackles, which can indicate developing pulmonary oedema (incipient heart failure). Women sometimes have persistent crackles in a localised area following previous surgery, and this should be recorded at the beginning of pregnancy so as not to be confusing later on. Sometimes women develop crackles as a result of poor lung expansion late in pregnancy, when the diaphragm is splinted by the enlarging uterus. Asking the woman to take several deep breaths and cough several times will usually cause such crackles to disappear. Crackles of pathological significance will persist despite such manoeuvres.

Any woman who complains of feeling suddenly less well, who develops 'funny turns' (any loss of consciousness is always significant in such women), a sudden increase in shortness of breath or new palpitations associated with other symptoms should always be assessed carefully by a cardiologist. An electrocardiogram is an important initial investigation. In tertiary centres it is usually possible to obtain an emergency echocardiogram 24/7. Arterial blood gas measurement can be informative, as can a chest X-ray, taken with screening of the fetus. If the woman complains of chest pain, it is useful to take blood immediately for measurement of troponin I levels and repeat the test 24 hours later to assess whether there has been any significant myocardial damage. In tertiary centres, an exercise treadmill test is the first non-invasive test of choice to investigate the possibility of coronary artery disease, assuming the patient is well enough. A myocardial perfusion scan or coronary angiography can be considered if symptoms continue or worsen despite treatment. Pulmonary embolism should also be considered and blood taken for measurement of d-dimer levels – if these are raised, anticoagulant treatment is probably the safest response. In doubtful cases, a ventilation/perfusion scan or computed tomography pulmonary angiography should be carried out, depending on local availability (bearing in mind that both expose the fetus to some radiation, particularly computed tomography scanning, although it is diagnostic in a higher proportion). Doppler examination of the leg vessels should be performed to identify any deep vein thrombosis. Dissection of the aorta should also be considered and may be detected on echocardiography, although magnetic resonance imaging is more sensitive, particularly for the thoracic aorta. Computed tomography scanning can also be used but exposes the fetus to a considerable radiation dose.

Management of a woman who develops new symptoms is dependent on the nature of the underlying lesion and the results of urgent investigations of cardiac function. It is not possible to give a brief account of the various management strategies which will be necessary, because they vary depending on the underlying lesion. Women with cyanotic heart disease, valvular disease, aortic dissections or arrhythmias require very different management, and many women will have an almost unique combination of lesions, requiring management tailored to their individual diagnosis. This is why an experienced cardiologist used to seeing pregnant women should always be involved in their care, especially in emergencies.

APPENDIX D

Pregnancy in women with heart disease: the typical patient journey

All women with heart disease should be assessed at the time of puberty (typically around the age of 12–15 years) by clinicians with expertise in the management of pregnancy complicated by heart disease. They should be given an estimate of their risks which is as accurate as possible, and this risk should be reassessed every five years (or more often if their condition deteriorates significantly). They should be advised whether specialist care from a high-risk pregnancy with heart disease team is advisable in the event of pregnancy. If so, they should be advised to see the appropriate high-risk team as soon as a pregnancy is confirmed, which will usually be by a urinary pregnancy test within two weeks of the missed period.

Women who present initially to their general practitioner or community/local hospital midwifery service, and give a history of heart disease should be referred promptly to an appropriate high-risk pregnancy and heart disease team.

At the initial assessment by the high-risk multidisciplinary team, a full clinical examination should be carried out and all recent investigations reviewed. Usually, an echocardiogram will be ordered to assess cardiac function. An electrocardiogram should be taken and kept in the notes for future reference, in the event that there is any change in cardiac status. The special antenatal notes should be started. The woman should be asked to carry her notes with her at all times, in case of any emergencies.

It is important to offer the woman a fetal nuchal translucency scan, as this is a significant indicator of recurrent cardiac disease in the fetus. It is usually carried out at 12 weeks of gestation. Once this scan has confirmed a viable fetus without obvious abnormalities, a standard fetal anomaly scan at approximately 20 weeks of gestation, and a fetal cardiac scan at approximately 22 weeks of gestation, should be organised.

Depending on her cardiac status, the woman should be seen by an appropriately experienced consultant obstetrician every two to four weeks until 20 weeks of gestation, then every two weeks until 24 weeks of gestation, and then weekly thereafter. Continuity of carer is of particular importance, because this makes it much easier to detect any deterioration in the woman's condition.

If the woman threatens to go into labour before 34 weeks of gestation, immediate assessment by the multidisciplinary team is important to assess the best management.

In pregnancies that are progressing satisfactorily, a multidisciplinary team assessment at 32–34 weeks of gestation is important to plan care around the time of delivery and to establish optimum management. The delivery plan proforma should be completed.

The woman should be given clear instructions about how to recognise the onset of labour. Once labour begins, she should immediately ring the labour ward to alert them that she is coming. She needs to make sure that they appreciate she is a cardiac patient so that they do not give her advice to wait at home, go for a bath, etc.

On arrival at the labour ward, the woman should make herself known immediately to the labour ward staff. They should inspect the delivery plan and take action accordingly. This is likely to include informing senior staff, usually consultants, of the woman's admission.

The majority of women with significant lesions will have epidural anaesthesia during labour, and a significant number will have an assisted vaginal delivery. All anaesthetics should be given by senior staff who are familiar with the delivery plan and have experience of pregnant women with cardiac disease.

Following delivery, the woman should be transferred to a high-dependency area where she can be monitored closely for anything between 12 and 48 hours. She should not be transferred to a normal labour ward until she has been reviewed by senior staff (preferably consultants) who can determine whether she will be safe in an area where monitoring will be less intensive.

Plans for discharge from the maternity unit should have been made antenatally. Before discharge, a check should be made that the woman has appropriate appointments for obstetric and cardiac follow-up and that she is aware of her contraceptive options.

At the postnatal check-up, the woman should be assessed for her recovery from giving birth. The woman's contraceptive plans should be reviewed in detail. Her cardiac function should be checked by a cardiologist, and arrangements made for cardiological follow-up.

APPENDIX E

Adapted antenatal record for use in women with heart disease

PREGNANCY DETAILS

Name: ..... Cardiac lesion: ..... S/B

EDD: Medications .....

Previous pregnancies: .....

Gestational age (weeks) .....

Birthweight (g) .....

Mode of delivery .....

DELIVERY PLAN:

Obstetrician .....

Cardiologist .....

Anaesthetist .....

Fetal echocardiogram .....

Date	Gestation	Shortness of breath	Palpitations	Other symptoms	Blood pressure	Pulse rate	Pulse rhythm	Murmur	Lung bases	Oedema	SFH (cm)	Presentation	5ths palp	FH	Urine	Next visit	Signature

APPENDIX F

Clinical management plan for delivery

Cardiac diagnosis……Please circle agreed plan

If admitted to LW, please inform	Consultant obstetrician on call **Yes / No** Obstetric SpR on call **senior / junior** Consultant anaesthetist on call **Yes / No** Anaesthetic SpR on call **senior / junior** Special midwifery team **Yes / No**	If advice is needed, please contact one of the following consultants via switchboard:
Mode of delivery	Elective LSCS / trial of vaginal delivery	
Elective LSCS (see anaesthetic sheet for anaesthetic details)	Prophylactic compression suture Syntocinon 2 units over 10–20 minutes Syntocinon low-dose infusion (8–12 mU/minute – see over for details) Anaesthetic technique…… Maternal monitoring……	Inform consultant on call if admitted in labour before scheduled date Epid/Spin/CSE/GA ECG/SaO_2/non-invasive BP/arterial line BP/ CVP
Vaginal delivery 1st stage Mx (see anaesthetic sheet for anaesthetic details)	TED stockings in labour/HDU chart Prophylactic antibiotics: If operative delivery / in all situations Epidural for analgesia…… Maternal monitoring…… Continuous EFM is recommended for all women with cardiac disease	Medications to be continued: …….. ● As soon as in established labour ● If and when requested ECG/SaO_2/non-invasive BP/arterial line BP/ CVP
Vaginal delivery 2nd stage Mx	Normal second stage Short second stage…… Elective assisted delivery only	Assist if not delivered in ……. minutes
Vaginal delivery 3rd stage Mx	Normal active Mx (oxytocin 5 i.u. IM and CCT, or 2 i.u. IV over 10 minutes) or Syntocinon infusion 8–12 mU/minutes …… (for details, see overleaf)	DO NOT GIVE ergometrine Continue …….. hours
Postdelivery	High-dependency unit **Yes / No** For …….... hours LMW heparin **Yes / No** Dose……Duration…… List medications to be given……..…… and continued for……...days/weeks Recommended post-natal stay ……days Cardiac review **Yes / No** weeks…… Contraceptive plans discussed **Yes / No**	

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.

Examples of clinical situations	Consider the following:
Spontaneous labour and recent thromboprophylaxis use (e.g. LMWH/warfarin) An epidural can be given more than 12 hours after prophylactic dose or more than 24 hours after therapeutic dose, or earlier at the discretion of the anaesthetist	Inform anaesthetist asap D/W consultant anaesthetist on call For additional advice, contact ……. …….. via switchboard, or obstetricians listed overleaf
Need for syntocinon augmentation in labour	Use double-strength syntocinon but halve rate to reduce total volume of fluids given (this decision needs to be taken at consultant level)
Syntocinon as prophylaxis against postpartum haemorrhage	Low-dose infusion (12 mU/minute): use either 5 i.u. in 50 ml at 7 ml per hour or 10 i.u. in 500 ml at 36 ml per hour Continue for 4 hours (longer if concerns)
Postpartum haemorrhage	Inform anaesthetic and obstetric consultants on call For uterotonic, misoprostol 600 micrograms rectally is preferred, but monitor for hyperpyrexia Avoid hemabate or high-dose syntocinon Consider use of compression suture Consider use of intrauterine balloon (antibiotic cover is recommended) Strict input/output charts to be maintained Consider central access or arterial monitoring
Preterm labour	Atosiban (tractocile) is the first-line Mx Do not use ritodrine or salbutamol
Pacemaker	Do not use unipolar diathermy Beware pacemaker in unusual places (e.g. abdominal wall) when performing caesarean section De-activate implantable defibrillators

Please seek advice from a consultant if there are concerns or if clarification is required on clinical management.

The RCOG is grateful to Ms Gubby Ayida FRCOG (Chelsea and Westminster Hospital) for permission to use this clinical management plan.

