Heart disease is the most common medical cause of maternal death in the UK and it is a major pregnancy problem around the world. This year, the Royal College of Obstetricians and Gynaecologists invited 26 of the world’s leading experts from the UK, Europe, the USA and Japan, to a study group, each contributing the latest available information in this vital area. This has resulted in a book of 24 chapters comprehensively detailing current knowledge, ranging from preconception counselling and contraception, through antenatal care of the mother and fetus, to supervision of delivery and the puerperium, and long term outcome. Specific conditions discussed include valve disease (and prostheses), ventricular disorders, arrhythmias, cardiomyopathies, ischaemic heart disease and endocarditis. The book ends with 53 consensus views on key issues, such as mode of delivery and emergency care, appropriate investigations and safe management. The editors have paid great attention to accuracy but also to readability. The fact that the contributors came together to check, refine and develop their chapters and the speed with which the book has been produced have resulted in a volume which is particularly valuable for updating clinical practice in this key area. This is a book that all high-risk obstetricians, and cardiologists and anaesthetists dealing with pregnant women with heart disease should read. Anyone with an interest in women’s health care will find something for them in these pages.
Heart Disease and Pregnancy
Since 1973 the Royal College of Obstetricians and Gynaecologists has regularly convened Study Groups to address important growth areas within obstetrics and gynaecology. An international group of eminent clinicians and scientists from various disciplines is invited to present the results of recent research and to take part in in-depth discussions. The resulting volume, containing enhanced versions of the papers presented, is published within a few months of the meeting and provides a summary of the subject that is both authoritative and up to date.

**SOME PREVIOUS STUDY GROUP PUBLICATIONS AVAILABLE**

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Heart Disease and Pregnancy

Edited by

Philip J Steer, Michael A Gatzoulis and Philip Baker
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**DECLARATION OF INTEREST**

All contributors to the Study Group were invited to make a specific Declaration of Interest in relation to the subject of the Study Group. This was undertaken and all contributors complied with this request. Helena Gardiner is a medical advisor to Echo UK, the fetal heart charity. Kate Harding has shares in GlaxoSmithKline. Martin Lupton is a consultant for the Roman Catholic Archdiocese of Westminster. Philip Baker’s department receives financial support from Pfizer UK for a clinical trial. Tommy's, the baby charity, contributes to the funding of the group and to his personal salary. He holds a patent for diagnostic testing for pre-eclampsia. Philip Steer is the Editor-in-Chief of BJOG – An International Journal of Obstetrics and Gynaecology. Steve Yentis was Honorary Secretary of the Obstetric Anaesthetists Association until May 2006.
Preface

In the Confidential Enquiry into Maternal Deaths in the United Kingdom 2000–2002, cardiac disease for the first time became the leading medical cause of death in relation to pregnancy. This prompted the Meetings Committee of the Royal College of Obstetricians and Gynaecologists in London to suggest heart disease and pregnancy as an important topic to be addressed as part of the regular series of Study Groups organised and funded by the College. This met with the approval of Philip Baker, Convenor of Study Groups, and so Philip Steer (obstetrician) and Michael Gatzoulis (cardiologist) set about bringing together a distinguished group of obstetricians, maternal medicine specialists, anaesthetists, obstetric physicians and cardiologists (both adult and paediatric) with an interest in care before, during and after pregnancy to consider this vitally important topic. A particular challenge was to gather together the limited amount of scientific data in an area of practice based largely on anecdote and relatively small case series, and from this restricted knowledge base to construct a consensus on issues of diagnosis and management. Accordingly, each participant was asked to prepare a presentation, on which the following chapters are based, on a specified aspect of the topic and subsequently to amend their chapter in the light of the discussions at the meeting, which was held on 13–15 February 2006 at the RCOG in London.

From the perspective of the participants, the meeting itself was a great success. There was an unusual sense of mission because all of those taking part are heavily involved in promoting effective care of women with heart disease who are or who wish to become pregnant, and are working hard to raise the profile of what is becoming almost a subspecialty in its own right. Some may question such a degree of specialisation but, with cardiovascular disease now becoming the leading cause of death in women generally, collaboration between cardiologists and obstetricians is only going to increase in this area in the future.

The chapters are laid out in a way that we hope readers will find intuitive, starting with prepregnancy counselling and contraception, moving through the antenatal period to delivery and the puerperium, and finishing with long-term outcome (about which distressingly little is as yet known – this should be an important focus for future research).

Finally, we list the consensus statements signed up to by all the members of the Study Group. There are a gratifyingly large number of these, which is a tribute to the flexibility and open-mindedness of the participants. It has to be admitted that most are based upon opinion rather than hard data. Nonetheless, we hope that readers will
find them a useful basis on which to consider building or modifying their own guidelines for practice, and upon which future more evidence-based guidelines can be constructed.

This volume, which has resulted from the meeting, is much more than the transcript of a fascinating meeting. Because of the way the meeting was designed, it also covers the clinical aspects of heart disease in pregnancy in a more comprehensive way than would have been possible had we produced a traditional meeting report. Indeed, we would like to see this volume as an initial attempt to produce a comprehensive text for those who take on the challenge of caring for this high-risk group of women. It brings together not only the data that we have available but also the considered opinions of many who see it as their life’s work to advance knowledge in this area. We thank the participants who gave their time freely to help make the meeting such a success and who have produced a series of outstanding chapters. Outcomes of pregnancy in women with heart disease will not always be good but we hope that the knowledge and opinions documented in this book will help to improve them in the future.

Philip J Steer
Michael A Gatzoulis
Philip Baker
Section 1

Prepregnancy counselling and contraception
Chapter 1

Preconceptual counselling for women with cardiac disease

Sarah Vause, Sara Thorne and Bernard Clarke

Introduction

The majority of women want to have children and women with heart disease are no exception. Complex heart disease is no bar to sexual activity. Most women with heart disease do have some awareness of the risks of pregnancy but their ideas are often inaccurate, ranging from overly optimistic to overly pessimistic. They may be equally poorly informed about the prognosis of their heart condition, even in the absence of pregnancy. Many doctors do not have a good understanding of the risks of pregnancy in women with heart disease and thus such women may be deprived of appropriate advice and counselling unless a specialist referral is made. Discussions with a cardiologist and/or an obstetric physician with a specialist interest in pregnancy and heart disease should begin in adolescence. These discussions should cover future pregnancies and their prevention, both to prevent accidental and possibly dangerous pregnancies and to allow them to come to terms with their future childbearing potential. They also need to be able to plan their families in the knowledge of their likely future health and life expectancy.

In the UK, the majority of women seen preconceptually by cardiologists and/or obstetricians will be women with congenital heart disease (CHD). This is because the incidence of CHD (0.8%) in pregnant women in the UK is higher than the incidence of acquired heart disease (0.1%). Furthermore, most women with CHD are already known to the cardiac services. Many women with acquired disease are recent immigrants and are unaware of their condition, which is an important reason why they are at particularly high risk. The diagnosis may be made only when they are pregnant and become symptomatic. For this reason, deaths in pregnancy from acquired heart disease outnumber those from CHD.

Components of preconceptual counselling

Preconceptual counselling should ideally:

- display attitudes and practices that value pregnant women, children and families and respect the diversity of people’s lives and experiences
- incorporate informed choice, thus encouraging women and men to understand health issues that may affect conception and pregnancy
encourage women and men to prepare actively for pregnancy, and enable them to be as healthy as possible

attempt to identify couples who are at increased risk of producing babies with a congenital abnormality and provide them with sufficient knowledge to make informed decisions.

These four components will be discussed below in relation to cardiac disease in pregnancy.

Valuing pregnant women, children and families and respecting diversity

Preconceptual counselling should display attitudes and practices that value pregnant women, children and families and respect the diversity of people’s lives and experiences. All women have a cultural context within a multicultural society. For some women, issues related to culture may need specific attention, including:

- their religious beliefs (particularly in relation to contraception and termination of pregnancy)
- the role of the partner and extended family in pregnancy decisions
- communication, where English is not the first language.

Assumptions are often made about the anticipated views of certain racial, cultural or religious groups and this may consciously or subconsciously affect the way in which doctors counsel women. Addressing these overtly helps one to compensate for any unintentional bias.

All people have a social and emotional context and when women and their partners seek advice this context must be considered. Their attitudes and expectations are likely to have been influenced by their previous experiences and those of their family. These may include:

- the anxieties of over-protective parents
- worries relating to their inability to embark on, or continue, a meaningful relationship if pregnancy is contraindicated
- wanting to fulfil their partner’s desire for a child, or guilt if they cannot do so
- ‘I’m lucky to be alive – am I pushing my luck or being greedy? Do I deserve a child?’

While it is important to explore and respect the context of her previous experiences, preconceptual counselling should promote the autonomy of the woman. It should enable her to determine her personal priorities and support her decision making.

Informed choice and understanding

Preconceptual counselling should incorporate informed choice, thus encouraging women and men to understand health issues that may affect conception and pregnancy. The counselling should provide information in a frank, honest and understandable way so as to give the woman a realistic estimate of both maternal and fetal risk and allow her to make an informed decision as to whether to embark on a pregnancy. The
counselling should include information on:

- the effects of cardiac disease on pregnancy, in terms of both maternal and fetal risks
- the effects of pregnancy on cardiac disease, including the risk of dying or long-term deterioration
- whether these effects will change with time or treatment
- the other options that are available, such as contraception, surrogacy or adoption
- the long-term outlook – a woman with a short life expectancy may feel that neither pregnancy nor surrogacy nor adoption is appropriate, as a child may then have to deal with the terminal illness and death of the mother

The risk of maternal death during pregnancy is often difficult to quantify but it is important to give the woman, her partner and her family as accurate an assessment of risk as is possible within our current state of knowledge. Although for the majority of women the risk will be less than 1%, for some conditions the risk may be considerably higher. Such high-risk cardiac conditions, where the risk is greater than 1% and in some cases up to 50%, include:

- any form of cyanotic CHD, for example Eisenmenger syndrome
- pulmonary hypertension
- poor systemic ventricular function, for example a systemic right ventricle
- severe left heart obstructive lesions, for example mitral and aortic stenosis
- Marfan syndrome, especially if the aortic root is dilated
- previous repair of a coarctation with a Dacron™ (polyethylene terephthalate) patch
- previous peripartum cardiomyopathy
- poor cardiac function for any reason at the time of conception.

The risks to the fetus include the following:

- intrauterine growth restriction with cyanotic heart disease
- iatrogenic prematurity
- fetal abnormality (typical risk of 3%, but this varies with the type of maternal lesion, and is also related to paternal lesions)
- teratogenesis, for example from warfarin or captopril
- fetal loss resulting from invasive prenatal testing.

For some complex conditions, there is little or no information available, either because of the rarity of the woman’s disease or because they represent a new cohort of survivors to adulthood with surgically modified disease. These women need a thorough assessment of their current cardiac status and appropriate advice based on how their cardiovascular system is likely to adapt to the physiological changes of pregnancy.

One successful pregnancy should not engender complacency. Some conditions, such as peripartum cardiomyopathy, have a high recurrence risk. Other conditions could worsen with age and in each subsequent pregnancy the risks would be higher.
Information about contraception and termination

Facilitating informed choice also means that doctors must provide information relating to the choice of not getting pregnant and thus appropriate contraceptive advice. Informed choice also means that the woman should be aware of the option of termination of pregnancy should she find herself unexpectedly pregnant after deciding that she was not planning to conceive. The assurance that clinicians will be non-judgemental and supportive of a decision to terminate a pregnancy is important. Open discussion of this option and provision of contact numbers to facilitate access to services reinforces that this is an option available to the woman. Termination of pregnancy in women with heart disease is not without risks, and it should be performed in a centre with appropriate anaesthetic and cardiac facilities.

Information about clinical management

During the preconceptual appointment the proposed plan of care for the pregnancy should be outlined. Women with significant cardiac disease should be managed in a centre with appropriate expertise, preferably in a joint obstetric–cardiac clinic, but for some women this may mean travelling long distances. Women should be made aware of how likely it would be that they would need admission antenatally, iatrogenic preterm delivery, lower segment caesarean section (LSCS) or high-dependency care in a hospital that may be many miles from home. However, for most cardiac conditions a normal vaginal delivery with good analgesia and a low threshold for forceps assistance is the safest mode of delivery for the mother, since it is associated with less blood loss and less rapid haemodynamic changes than caesarean section. The few maternal cardiac indications for delivery by caesarean section include Marfan syndrome, aortic aneurysm of any cause, and an acutely unwell mother.

Information in an appropriate language

If the woman and her doctor do not speak the same language, a professional interpreter should be used. Interpreters from within the family, including the husband, should not be used as in the family's desire to help the woman have a successful pregnancy the risks may not be accurately relayed to her.

Preparing for pregnancy

Preconceptual counselling encourages women and men to prepare actively for pregnancy and enables them to be as healthy as possible. The consultations provide the ideal opportunity to minimise risk and optimise cardiac function before pregnancy:

- valvotomy by catheter or surgical intervention before pregnancy – if valve replacement is performed the choice of the type of valve used may be influenced by the desire for a further pregnancy; the use of tissue valves obviates the need for anticoagulation during pregnancy
- treatment of arrhythmias (surgical or medical)
- treatment of underlying medical conditions, such as hypertension or diabetes
- avoidance of teratogens – medication may need to be changed before pregnancy
- discussion re anticoagulation – women using warfarin need to be aware of its teratogenic potential and the risk of fetal intracranial haemorrhage, and they
should understand the need for conversion to heparin once pregnancy is confirmed. Contact numbers should be provided to facilitate this as early as possible in pregnancy. The involvement of haematologists will enable appropriate dosing and monitoring. For women with mechanical valves the risks of warfarin versus the risks of heparin should be discussed with them to enable them to make an informed choice.

- dental treatment – women with complex heart disease may need to be referred to a tertiary dental hospital for dental care; it is better for any dental problems to be resolved before pregnancy
- timing of pregnancy – for those with a systemic right ventricle or univentricular heart, pregnancy is likely to be tolerated better when the woman is in her 20s rather than her late 30s. A woman’s life may not allow her the luxury of making this choice but other women may have a choice and should be discouraged from purposely delaying pregnancy because of considerations such as a career.
- contraception – until the above cardiac problems have been appropriately addressed, the provision of appropriate contraception is important
- general pre-pregnancy advice should not be forgotten, for example folic acid
- provision of phone numbers to facilitate prompt contact and reassessment once pregnancy is confirmed.

Women undergoing assisted conception often have additional risk factors such as increased age, and the risk of ovarian hyperstimulation and multiple pregnancy with a concomitant increase in the risk of pre-eclampsia. These conditions can compound the risk of heart disease. In women undergoing assisted conception, it is important that precautions should be taken to avoid hyperstimulated cycles and to minimise the chance of multiple pregnancy by only carrying out single-embryo replacements during in vitro fertilisation (IVF) cycles.

**Risk of congenital abnormality**

Preconceptual counselling attempts to identify couples who are at increased risk of producing babies with a malformation, and should provide them with sufficient knowledge to make informed decisions. Many couples have worries about the recurrence risk to their unborn baby. For the majority of women with CHD, with no family history and no chromosomal abnormality, the risk of recurrence of CHD in the fetus is around 3%. Prenatal fetal echocardiography can be arranged and most couples can be reassured that the most likely outcome is a healthy baby.

For women with a family history of heart disease, or other features to suggest an underlying genetic or chromosomal problem, the preconceptual appointment offers the opportunity to refer a woman to the clinical geneticist or to arrange karyotyping if this has not previously been done. Karyotyping may detect balanced translocations or the 22q11 deletion. While the woman may have previously been seen by a geneticist, she may welcome the opportunity to discuss the risks to the fetus again once she begins to contemplate pregnancy.

Preconceptual care should also include a discussion of the various prenatal tests available, their risks and limitations, the timing of the tests and the way in which they are performed. Information about how to access these tests, including contact numbers, should be provided. Discussion should also include the options available, including termination, should the fetus be found to be affected.
For other women with conditions such as Marfan syndrome the preconceptual appointment offers the opportunity to discuss the recurrence risk, and the need for appropriate follow-up of the infant postnatally to establish a diagnosis and, if needed, to implement long-term surveillance.

Conclusion

Successful preconceptual counselling will empower a woman with cardiac disease to make informed choices relating to pregnancy by providing non-directive counselling and access to the appropriate multidisciplinary specialised services. Optimising her health before pregnancy will improve the likelihood of a successful pregnancy outcome.

References