Maternal cardiovascular medicine: towards better care for pregnant women with heart disease

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Abstract

Cardiac disease has emerged as the leading cause of maternal death during pregnancy in the UK. Its incidence has been rising in the past two decades, largely due to increasing mortality from acquired heart disease, which currently exceeds mortality from congenital heart disease. According to the Confidential Enquiry, better care could have altered the course of 40% of the deaths from cardiac causes. Management of these patients is critical, because any maternal complication has major impacts on surviving children. Improvements in maternal cardiovascular medicine require concerted efforts through interdisciplinary collaboration of all specialties caring for pregnant cardiac patients. One important area for improvement is on how to identify and evaluate those at highest risks of pregnancy-related cardiac complications. Most assessments and clinical guidelines for the management of pregnant women with heart disease have been based on retrospective lesion-specific information. Direct evaluation of cardiac function during pregnancy may add further information, improving the cardiac care we can provide to individual pregnant cardiac patients. More research in this area is urgently needed. We also propose that improvement in training, research and exposure to the subspecialty of maternal cardiovascular medicine is needed to continue to raise standards of care for this patient population.

Key words: pregnancy, maternal mortality, maternal cardiovascular medicine, cardiac evaluation, risk stratification.

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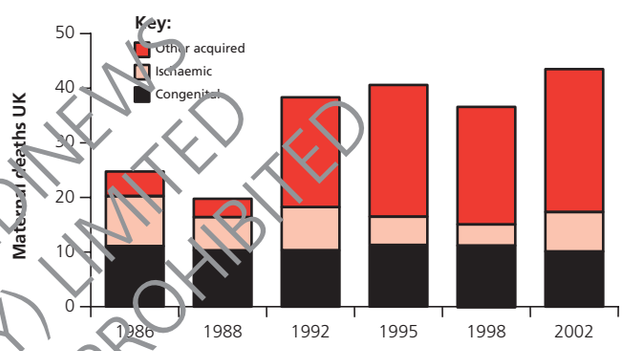
Figure 1. Maternal mortality rates from cardiac causes of death in the UK 1985–2002



Adapted from Lewis G[4]

Introduction

Pregnancy and childbirth are natural life events and any related death must be considered to be 'one too many'. Although relatively uncommon in the UK, maternal death, whenever it occurs, is a very tragic event with surviving children becoming motherless. The importance of this issue is emphasised by the World Health Organisation's World Health Report: *Make every mother and child count*.[1] The risk of maternal death is about one in 10,000 pregnancies in the UK. This compares to more than one in 100 pregnancies in parts of Africa,[2] and as high as one in 11 for women in Chad.[3] Nevertheless, in the UK, heart disease still poses major risks to pregnant mothers and continues to be a leading cause of maternal mortality.[4]

Confidential enquiry into maternal deaths

Over 50 years ago, a national system was set up for reporting regional and national maternal deaths in the UK. A series of triennial reports was instituted to publish the findings and associated recommendations for improving practice to reduce maternal mortality. The most recently published Confidential Enquiry into Maternal Deaths (CEMD), *Why mothers die 2000–2002*, reported that heart disease has now become the leading indirect cause of maternal deaths (i.e. not directly related or caused by complications of pregnancy itself, but due to pre-existing condi-

tions made worse by the pregnancy) with 44 cases in the UK in 2000–2002.[4] Nine of the maternal deaths in 2000–2002 were due to congenital heart disease and this incidence has been relatively stable over the past two decades (figure 1). The rest of the deaths (35) were due to acquired heart disease. The progressive rise in cardiac deaths (figure 1) is more related to acquired disease, such as myocardial infarction and cardiomyopathy. These two conditions were responsible for eight deaths each in the last report.[4] Acquired heart disease during pregnancy, especially cardiomyopathies, is becoming more common in western countries;[4,5] partly due to enhanced awareness, diagnoses, case-ascertainment and rising maternal age in recent decades.[6]

High maternal mortality in pregnant cardiac patients has prompted the assertion that women with cardiovascular disease should avoid or discontinue pregnancy. This is partly based on conventional cardiological wisdom that serious maternal morbidity, such as heart failure, thrombo-embolism and arrhythmias, are more common in women with pre-existing heart disease.[7] But despite these risks, it is now accepted that pregnancy in most women with heart disease usually has a favourable maternal and fetal outcome.[8] Avila pointed out that the long-standing notion that pregnancy is contraindicated in cardiac patients is no longer tenable today.[9] There is a need to attain the optimum balance between avoiding tragedy, whilst supporting the majority of women to achieve their basic human right of a safe and successful pregnancy and labour, with the ultimate delivery of a healthy baby.

Improving management of maternal cardiac disease

Of greatest concern is the fact the Confidential Enquiry assessors considered that some degree of substandard care was present in 40% of the deaths. To minimise these deaths, there are a number of initiatives that urgently need to be considered, debated and put into place. These should include education, training, resources, increased facilities, more research and a greater understanding of the key problems. A first step is for clinicians to be aware and up-to-date about what is at stake for each pregnant patient from a cardiological point of view.

Normal gestation is accompanied by maternal cardiovascular adaptations, which include increased cardiac output and tachycardia. Pregnancy itself can therefore be viewed as a process where protracted, finite cardiac stress is brought about by the need to supply extra circulation to the gravid tissues. These physiological cardiovascular adaptations can potentially worsen the prognosis in women whose pregnancy is complicated by heart disease.[10] The cardiovascular system is then further stressed by the physical strain of labour or Caesarean section.[11] During each uterine contraction, cardiac output is increased by 34% and blood pressure by 12%.[12] Knowing what constitutes high risk and when to refer to a speciality clinic are key to managing women with congenital or acquired heart lesions.[13]

Occult heart disease may be present in a young mother,[14] and may present with non-specific symptoms such as palpitations, breathlessness, fatigue or oedema, all of which can mimic cardiac symptoms but are common in a normal pregnancy.[15] It is challenging for all healthcare professionals dealing with pregnant women to differentiate the benign symptoms and signs from those heralding serious cardiac compromise. A high index of suspicion for cardiac diseases is essential to identify those at risk. Once these patients are referred for a cardiological opinion, there is a need for cardiologists to develop a systematic approach to their evaluation.

Current methods of evaluations

Established methods for evaluating pregnant cardiac patients are broadly based on three types of evidence:

- lesion-specific retrospective information
- cumulative clinician experience, and
- multivariate modelling.

These provide probabilistic information to guide clinicians in estimating the likelihood of cardiovascular complications during pregnancy and labour in individual patients. The more accurately clinicians can determine the probability of risks, the less likely it will be that they will advocate the negative mentality that 'cardiovascular risks equate contraindication to pregnancy', and shift towards a more positive attitude of 'exploring means to minimise the risks'. Ideally, these considerations should be commenced during pre-pregnancy consultations, but continued throughout pregnancy and the post-partum period.

Lesion-specific information

The traditional medical school teaching of heart disease in pregnancy is lesion-specific. For example, information gathered on mitral stenosis does not generally apply to pulmonary regurgitation. This approach is also used in most review articles and guidelines covering maternal cardiovascular disease.[14,16-22] The current evidence base is mainly built upon experiences from retrospective case reports and series in different locations with diverse resources and skill-mix. This is partly due to the fact that the issues surrounding these pregnant patients cannot be answered by randomised controlled trials, given the variability in anatomy and physiology in women with heart disease and the inappropriateness of randomisation in this setting.[23]

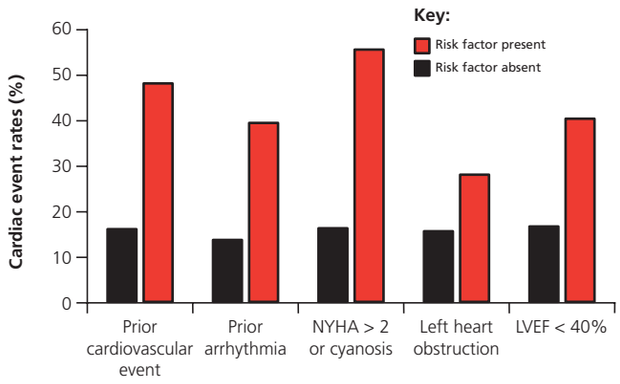
Cumulative clinician experience

As there is often a limited amount of firm evidence in this area, decisions are often based on intuitive expertise. Decisions may involve value and probability judgements;[24] i.e. an assessment of how likely it is that an individual woman with heart disease may suffer an adverse maternal or fetal event, as well as how terrible these events would be. It must be borne in mind that anecdotal experiences may sometimes disproportionately amplify tragic experiences, leading to an over-cautious approach being adopted and avoidance or termination of pregnancy advised. Subjective perspectives need to be balanced with objective evidence to reach a more dispassionate management plan.

Multivariate modelling

Factors that place the mother and fetus at risk can be identified before pregnancy. This allows the development of patient-spe-

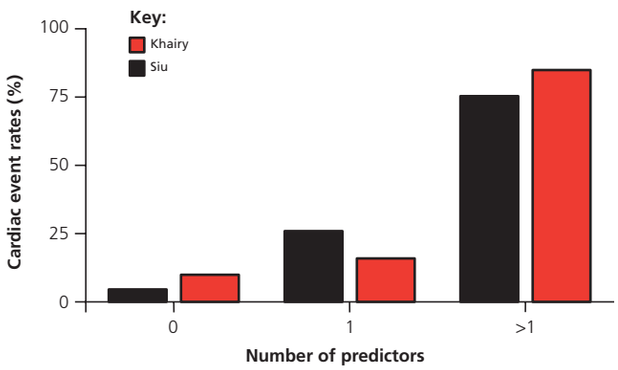
Figure 2. Independent predictors of cardiac events



Key: NYHA = New York Heart Association; LVEF = left ventricular ejection fraction

Adapted from Sermer[10] (with data from Siu[25])

Figure 3. Expected cardiac event rate based on risk index scoring system



Adapted from Sermer[10] and Khairy[27] (with data from Siu[25])

cific management plans.[10] With this objective, Siu *et al.*[25] conducted a large (276 completed pregnancies) retrospective, multicentre study to identify risk factors that predicted cardiac events in pregnancy (figure 2). A cardiac event was defined as cardiac death, stroke, pulmonary oedema, or arrhythmia. These risk factors were then validated through a larger (599 completed pregnancies) prospective multi-centre study.[26] Through multivariate analyses, they found the following independent risk factors:

- poor functional status (New York Heart Association [NYHA] class III or IV) or cyanosis
- left ventricular systolic dysfunction (ejection fraction [EF] < 40%)
- left heart obstruction (mitral valve area < 2.0 cm^2, aortic valve area < 1.5 cm^2 or peak left ventricular outflow tract gradient > 30 mmHg)
- a cardiac event (arrhythmia, stroke, transient ischaemic event, or pulmonary oedema) before pregnancy but since a prior cardiac surgical procedure.

A risk index scoring system was developed (figure 3)[26]:

- a woman with heart disease but no other risk factors has a 5% likelihood of a cardiac event during pregnancy
- her risk increases to 25% with one risk factor and
- her risk increases to 75% with more than one risk factor.

Using this system, it became possible to identify women at higher risks of pregnancy-related cardiac and neonatal complications. This is important for the counselling and management of this population.[10] Although this broad risk-stratifying approach represents a major step forward, a greater refinement of the risk-stratifying capability would be advantageous to clinicians. For example, NYHA classes III-IV encompass a fairly wide range of cardiac functional capacity from those requiring cardiac transplantation to those who can lead a reasonably normal life. With institution of appropriate therapy, some of these patients may improve to NYHA classes I or II. Similarly, cardiologists have many heart failure patients with an EF < 40% who can lead an active life with no need for diuretic therapy, while some patients with an EF > 40% are severely limited.

For other centres to adopt these risk scores, it is necessary to make additional assumptions that the characteristics of the patient population, the health care facilities, and the skill and expertise of the healthcare professionals are similar to those in Toronto in the late 1990s where and when this study was conducted. In particular, this study predated the widespread use of beta blockers in the treatment of heart failure. A recent attempt at validating these scores by Khairy *et al.* in a cohort of Boston patients with congenital heart diseases revealed rather different cardiac event rates, while identifying other new independent risk factors (figure 3).[27]

Direct evaluation of cardiac function

Since pregnancy, labour and the post-partum period constitute major stresses on the cardiovascular system, it is logical to evaluate whether the diseased heart of pregnant patients can cope with these stresses. It has been claimed that "any patient who reaches functional class III or IV during pregnancy is at high risk whatever the underlying condition, as this means that there is no remaining cardiovascular reserve".[16] However, as alluded to above, the subjective elements in attributing NYHA class mean there are inevitable uncertainties and misjudgements which require objective verification. Assessment of cardiac function by parameters including NYHA functional class, exercise capacity and peak oxygen consumption (peak VO_2) are useful as indirect indicators of cardiac (dys)function,[28] since the obligatory weight gain as gestation progresses leads to the development of exercise intolerance and a decline in functionality. These indirect indicators of cardiac function are subject to non-cardiac influences and thus become non-specific for cardiac compromise. There is therefore a need to evaluate cardiac function directly, taking into

account both the flow- and pressure-generating capacity of the heart.[28-29] Such cardiovascular physiological information is currently unavailable even in healthy subjects and is urgently needed to ultimately improve the cardiac care we can provide to pregnant patients.

Need for interdisciplinary collaborative work

A multidisciplinary health team approach is beneficial to both patients and health carers in the following situations: complex health problems, diseases where speed of management is critical, and in very emotionally challenging areas. Heart disease in pregnancy fits all of these criteria and also encompasses diverse specialists including obstetricians, feto-maternal medicine specialists, cardiologists, anaesthetists, midwives and primary care physicians. We believe that an interdisciplinary collaboration between patients, their families and these specialists is needed to deliver the ideal management for patients. It may not be an exaggeration to state that the weakest link in this chain may well lie in the lack of cardiology specialists caring for acquired cardiovascular diseases in pregnant patients. To address the root of this shortfall requires a rethinking of the contents of cardiological training to include more structured and systematic programmes.

Training issues

Heart disease in pregnancy is part of the general cardiology training curriculum[30] although this is usually not structured. It is assumed that the knowledge will be picked up during traineeship, which usually means a trainee reviewing a few pregnant women while on-call. Such informal arrangements leads to most specialist registrars in cardiology not being adequately exposed to maternal cardiovascular disease. We believe it is time to raise the profile of maternal cardiovascular medicine in the curriculum as a start to addressing the substandard care that was highlighted in the *Why mothers die* report.

More research needed in this area

Maternal cardiovascular medicine is currently a fringe subspeciality. Although there are many dedicated, enthusiastic and interested cardiologists, adult congenital heart disease specialists and maternal fetal medicine specialists carrying out vital work, there is currently no unifying body. Research is carried out in individual centres, with very limited funding. We feel it is time to increase awareness of maternal cardiovascular medicine amongst the funding bodies to set up much needed research at a national level. National and international cardiology congresses should be encouraged to include suitable abstract submission categories for this subspecialist subject, in order to reverse the current low profile of maternal cardiovascular medicine. Denying practising cardiologists exposure to these abstracts only serves to propagate disinterest in this important subject area.

Conclusion

Maternal cardiac disease is usually associated with a favourable outcome, yet cardiac disease remains a leading cause of the tragedy of maternal mortality. Deaths from acquired heart dis-

Key messages

- Cardiac disease is the leading cause of maternal mortality during pregnancy in the UK
- Substandard care was deemed to contribute to 40% of maternal deaths from cardiac causes in the latest Confidential Enquiry
- Interdisciplinary collaboration is essential to provide better care for these patients
- Improvements in training and research are vital to advance the subspecialty of maternal cardiovascular medicine

ease, in particular, appear to be increasing in the past two decades. Women with pre-existing cardiac disease are at higher risk and methods to predict poor outcome have been developed, but the tools for risk stratification need to be further refined in order to enhance the care that can be provided to these pregnant mothers. Developments in research, training and interdisciplinary collaboration are needed to raise the profile of maternal cardiovascular medicine and improve the outcome for future pregnant mothers with cardiovascular disease.

Conflict of interest

None declared.

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