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Review

Preconception counselling of the patient with systemic ventricular dysfunction

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ABSTRACT

With improvements in paediatric and adult cardiac care there are now large populations of young women with stable but significant left ventricular dysfunction. These women should all have access to expert multi-disciplinary pre-conception counselling as myocardial disease is a leading cause of maternal morbidity and mortality. This paper discussed the data that is available to the team delivering this counselling and also highlights the areas of uncertainty. Risk scoring systems and investigative tools are all discussed and practical suggestions are made regarding the specific issues that need to be raised in comprehensive counselling. The aim of effective counselling is to empower women to make informed decisions about their plans to start a family and to optimise care when a pregnancy occurs.

1. Introduction

With improvements in paediatric and adult cardiac care there are now large populations of young women with stable but significant left ventricular dysfunction. Many of these patients are well controlled on medical therapy and are leading lives that are difficult to differentiate from the normal population. The desire to have a family is part of that normality. These women do not, however, have a normal outcome during pregnancy and access to detailed pre-conception counselling is essential for all such women of child bearing age. This should also include women in their 40s, and even 50s, who are considering assisted fertility techniques.

This article aims to review the data available to the clinician providing such pre-pregnancy advice. It will also highlight the limitations in our current knowledge base and the areas of clinical uncertainty. This article will focus on systolic dysfunction of the systemic ventricle including those with systemic right ventricles in the context of transposition.

2. Why is pre-Conception Counselling Necessary?

Myocardial disease was the leading cause of maternal cardiac death in the latest UK Confidential Enquiry into Maternal Death [1]. An even more sizable issue is that of severe maternal morbidity as highlighted by the Netherlands National Cohort Study [2]. This is a registry covering all of the country's 98 maternity units serving a population of 17 million. In this study cardiomyopathy was a major cause of cardiovascular events significant enough to trigger emergency admission to hospital (Fig. 1).

Pregnancy is a challenge to a patient with impaired ventricular function. Increases in blood volume, elevations of heart rate and changes in vascular resistances are all components of the normal adaptive processes during pregnancy [3]. Increased tendency towards thrombosis and arrhythmia is also present. Pregnancy can also bring additional challenges including the need for invasive procedures, the need for vasoactive medication and pregnancy related complications such as pre-eclampsia. In a woman with a finely balanced but "vulnerable" ventricle the ability to increase cardiac output may be significantly blunted.

3. Tools for Stratifying Maternal Risk

There are several well recognised maternal scoring systems for cardiac disease. One of the original maternal cardiac studies was the CARPREG study from Canada [4]. In the CARPREG risk score points are awarded for various risk factors — this includes a history of a prior cardiac event (such as heart failure) and a pre-pregnancy reduction in systemic ventricular dysfunction of less than 40%. New York Heart Association (NYHA) assessment of functional class is also a risk factor if the patient is highly symptomatic (NYHA class of >II). The Zahara scoring system had similar predictors of outcome but also included the use of cardiac medication pre-pregnancy and the presence of severe atrioventricular valve regurgitation [5].

An alternative scoring system with more comprehensive application is the use of World Health Organisation (WHO) risk groups [6]. In the WHO classification patients with moderate ventricular dysfunction

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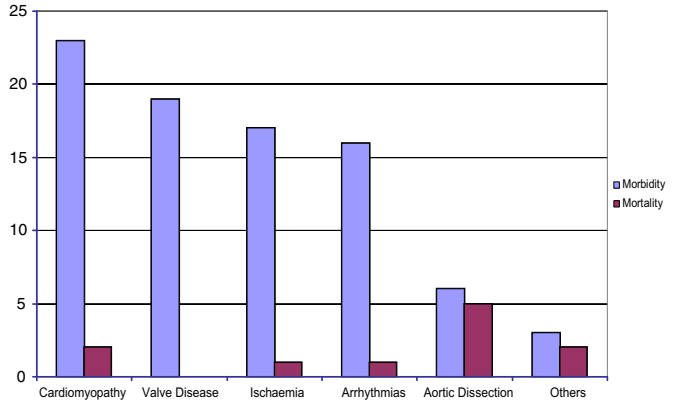

Fig. 1. Maternal cardiac morbidity and mortality per 100,000 deliveries. Adapted from the LEMMoN-Study [2].

would be in risk class III. This group is said to have a "Significantly increased risk of maternal mortality or severe morbidity". The recommendations then suggest that such women are cared for in a "specialist multi-disciplinary cardiac obstetric unit". More recently the European Society of Cardiology commissioned a registry study to assess actual maternal outcomes in these WHO groups. In this subsequent study patients in WHO class III had a maternal mortality of 1.5% and a risk of heart failure of 19%. Overall the risk of hospitalisation due to cardiac issues was 36% [7].

4. Cardiomyopathy

Cardiomyopathy was the commonest cause of morbidity from the Dutch registry [2]. In the latest UK confidential enquiry 13 women died over a 3 year period [1]. Of these 13 deaths, 9 were either suspected or proven peripartum cardiomyopathy. Peripartum cardiomyopathy (PPCM) is a rare complication of pregnancy and is a diagnosis of exclusion. It has four key features: 1 — development of heart failure in the last month of pregnancy or within five months post-delivery; 2 — absence of another identifiable cause for heart failure; 3 — absence of underlying heart disease prior to the last month of pregnancy; and 4 — evidence of left ventricular systolic dysfunction. There are many known risk factors including maternal age, obesity, hypertension, multiparity, multiple gestation, beta-agonist use and black race [8].

In women of child bearing age seeking pre-conception counselling the most common causes of ventricular dysfunction in a Western country are idiopathic and familial dilated cardiomyopathy (DCM), previous myocarditis, previous chemotherapeutic agent exposure, previous PPCM and congenital heart disease.

Treatment for all types of myocardial dysfunction during pregnancy is improving with a case fatality rate of only 10% [2]. Standard treatment includes diuretics, beta-blockers and occasionally hydralazine and nitrates. Angiotensin converting enzyme (ACE) inhibitors should be started post-natally. When there is acute severe heart failure, inotropic support, mechanical support (balloon pump or ventricular assist device) and transplantation may all be required. Delivery is indicated if cardiac failure cannot be treated adequately. The other specific treatment for PPCM is the dopamine antagonist bromocriptine and early data suggests that this may significantly improve maternal outcome [9].

As with all other cardiac conditions presenting in pregnancy early diagnosis is crucial. The onset of new asthma in pregnancy (particular in the third trimester or just following delivery) should be viewed with suspicion and there should be a low threshold for checking a brain natriuretic peptide (BNP) and performing an echocardiogram.

5. Other Risk Factors to Consider

The traditional pregnancy risk scores are a good starting place when counselling a woman with ventricular dysfunction. However they are not nuanced or comprehensive enough to be the only source of risk stratification. There are several other important considerations that are missing from the scoring systems. For example the aetiology of cardiomyopathy is an important determinant of outcome in the non-pregnant patient. The international childhood and adolescent cardiomyopathy registries show differing mortality for different sub-types of DCM. Those with neuromuscular-related and idiopathic cardiomyopathies have a poorer prognosis than those with underlying metabolic related DCM [10]. In addition the pre-conception trajectory of disease is likely to be important. When looking at two patients with an identical ejection fraction the one with a stable ejection fraction is likely to do better than the one with a steadily declining function.

6. Can we do Better Than Ejection Fraction?

Traditional pregnancy risk scores use ejection fraction as the assessment of ventricular function. More recently other parameters have been shown to be informative. In patients with previous PPCM, contractile reserve (from stress echo) has been shown to be impaired when compared to a control population. This is true even in those who had appeared to have a fully recovered ventricle. In 2010 Fett et al. used contractile reserve to predict recurrence of PPCM in subsequent pregnancies. In their study there were two interesting sub-groups. Both of these groups had an ejection fraction of greater than 55% — one had a normal contractile reserve; the other had an impaired reserve. PPCM did not recur in those with a normal reserve but did recur in 23% of those with an impaired reserve [11].

7. Newer Tools for Stratifying Risk

More recently exercise stress testing has been used as a part of comprehensive pre-conception counselling. These studies were not specific for DCM but were in more heterogeneous groups of women with heart disease. Peak heart rate and peak oxygen uptake are both higher in patients who go on to have a pregnancy without a maternal cardiac event [12]. In a separate study chronotropic response was shown to be associated with a reduced risk of maternal events during pregnancy [13].

8. Biomarkers

Several studies have looked at BNP (and N-terminal pro-BNP) during normal pregnancy. This stays in the region of 15–20 pg/L throughout pregnancy although it may increase in the presence of pre-eclampsia. In women with heart disease BNP is significantly elevated in those who have maternal cardiac events [14]. BNP has also been used to predict outcome when measured at 20 weeks [15]. In these studies an NT-proBNP of greater than 100 pg/mL is often associated with a maternal cardiac event during pregnancy. It is unclear if a pre-conception BNP also predicts outcome but this is likely. Many units now use serial measurements of BNP to monitor a woman's progress during pregnancy (Fig. 2).

9. Assessment on or off Therapy?

One of the unanswered questions is whether women should be assessed (pre-conception) on or off therapy. Several heart failure medications must be discontinued in pregnancy (such as ACE inhibitors or spironolactone) due to their associated teratogenicity. It is likely that discontinuing these drugs will have a deleterious impact on ventricular function. Our current practice is to perform the first assessment on therapy. If the woman is adamant that she wishes to have a pregnancy