Editorials

Congenital Heart Disease and Pregnancy. What Do We Know in 2008?
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Cardiac disease is the leading medical cause of material death during pregnancy in the United Kingdom.1 Due to recent advances in cardiac surgery and congenital heart disease, the butter is now the most common substrate of heart disease in pregnancy in the western world.2 The combination of cardiac disease and pregnancy carry mortality and morbidity risks such as heart failure, thromboembolism, and cardiac arrhythmia. Furthermore, fetal and neonatal adverse events, including intruterine growth restriction, premature birth, intracranial haemorrhage, and fetal loss are relatively common. Prepregnancy counselling (Table), including advice on contraception, and optimal care during pregnancy is, thus, becoming a major topic in current cardiologic and obstetric practice.

The first task is to assess the risks of pregnancy for the mother. Pregnancy is associated with profound changes in peripheral resistance, cardiac output, and blood volume, in order to provide appropriate uterine blood flow. In the first trimester of pregnancy, the blood pressure falls secondary to a drop in peripheral vascular resistance. Thereafter, plasma volume increases by 25%, and this change in volume is associated with an accelerated heart rate and a 50% increase in cardiac output. Blood pressure starts to rise in the beginning of the third trimester. Structural changes to the heart and great vessels also occur and include myocardial hypertrophy, chamber enlargement, and valvular regurgitation.3-11 These major cardiovascular changes may, in women with cardiac disease, be poorly tolerated and precipitate heart failure and clinical decompensation. Proarrhythmic effects and an increased likelihood of thrombosis could also compromise these patients.12

Labour and delivery is a particularly risky period. Their timing is crucial to balance maternal and neonatal morbidity, and mortality risks. A clear management plan should be established in advance and be communicated to all parties involved, including the patient. In general, vaginal delivery under epidural anaesthesia is preferable; it carries a lower risk of complications for both the mother and the fetus compared to caesarean section, namely lower risk of haemorrhage, infection, and thrombosis. But every case should be considered individually, taking into account not only obstetric and cardiac factors but also availability of the multidisciplinary team required. Large units are more likely to offer round the clock coverage, compared to smaller units; the latter may, therefore, need to utilize more caesarean section for scheduling purposes.

The risk of maternal death during pregnancy varies and has been reported to be as high as 30%-50% for patients with pulmonary hypertension.13,14 In contrast, mortality and morbidity risks are very small in patients with mild or repaired lesions and, in such cases, appropriate reassurance should be provided and local obstetric and cardiac care should suffice. While Siu et al5,6 and a recent meta-analysis from the Netherlands15 have provided important data on maternal morbidity and mortality, when it comes to the individual patient with congenital heart disease other features than the lesion itself, previous surgery and generic cardiac risk factors such as age, physical status, and smoking play also a role (Table 1).

The essence of preconception counselling is that it needs to be multidisciplinary, in good time (ideally in teenage years) and the limitation of existing data that guide us, the physicians, need to be discussed with the patient and the family. Furthermore, the uncertainty of the potential adverse effects of pregnancy on ventricular function needs to be acknowledged.16 Young female patients with valvular lesions should be taught not to accept metallic prostheses, before previous specialist consultation, as the need for anticoagulation will complicate significantly future pregnancy and increase maternal or neonatal risks (depending on the anticoagulation chosen).17 Nevertheless, when the

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### TABLE 1. Preconception Counselling for Women With Congenital Heart Disease

<table>
<thead>
<tr>
<th>Lesion Specific Risks</th>
<th>Low (1:1000-1:100)</th>
<th>Moderate (1%-2%)</th>
<th>High (&gt;2%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventricular septal defect</td>
<td>Mitral stenosis</td>
<td>Marfan syndrome with significant aortic root/AV involvement</td>
<td></td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>Aortic stenosis</td>
<td>Eisenmenger syndrome</td>
<td></td>
</tr>
<tr>
<td>Coarctation repaired</td>
<td>Systemic RV (TGA after atrial switch procedure or ccTGA)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>Fontan-type circulation</td>
<td>Cyanosis without PAH</td>
<td></td>
</tr>
</tbody>
</table>

**Generic Cardiac Risks**

- Poor functional class before pregnancy (NYHA >II) or cyanosis
- Impaired systemic ventricular function (EF <40%)
- Left heart obstruction (MV area <2 cm², AV area <1.5 cm², LV outflow tract peak blood pressure gradient >30 mm Hg before pregnancy)
- Preconception history of adverse cardiac events such as asymptomatic arrhythmia, stroke, transient ischaemic attack, and pulmonary oedema

**Other Risks**

- Age <18 and >35
- Physical deconditioning
- Smoking

**Fetal Risks (of Miscarriage, IUGR, etc)**

| Maternal NYHA functional class >II or cyanosis |
| Treatment with anticoagulants during pregnancy |
| Maternal smoking |
| Multiple gestation |
| Left heart obstruction |

**Risk of Transmission of CHD**

<table>
<thead>
<tr>
<th>Mother Affected</th>
<th>Father Affected</th>
</tr>
</thead>
<tbody>
<tr>
<td>11.6</td>
<td>4.3</td>
</tr>
<tr>
<td>8.0</td>
<td>3.8</td>
</tr>
<tr>
<td>6.3</td>
<td>3.0</td>
</tr>
<tr>
<td>6.0</td>
<td>3.6</td>
</tr>
<tr>
<td>5.3</td>
<td>3.5</td>
</tr>
<tr>
<td>2.0</td>
<td>1.4</td>
</tr>
</tbody>
</table>

**Maternal and Paternal Survival Prospects**

- Should be discussed: Parental life expectancy, Risk of premature death or disability, Need for major surgery

**Levels and Intensity of Health Care Required**

- **Pregnancy**
  - Low Risk Patients
    - Local hospital
    - 1 visit at ACHD clinic (clarify issues/make recommendations)
  - Moderate and High Risk Patients
    - Tertiary hospital
    - Multidisciplinary approach “Joint Cardiac Obstetric Clinic”

- **Delivery and Postpartum**
  - Vaginal delivery, epidural analgesia, and left lateral decubitus position
  - Only few cardiac indications for caesarea section (which is associated with rapid haemodynamic changes/more blood loss and higher risk of infection)
  - Advice level of monitoring required (ie, arterial lines, oxygen saturation, etc)
Preconception Counselling for Women With Congenital Heart Disease (Continued)

Effective Contraception and Termination of Pregnancy

**Contraception**

Timely contraceptive counselling
Several contraceptive methods suitable, recommend progestogen-only methods ie, Mirena IUS (intrauterine device), Implanon (subdermal implant)

**Termination of Pregnancy**

First trimester preferably
Facilities should be readily available where pregnancy places a woman’s life at risk

**References**