

## Editorial

# Family planning requirements of adults with congenital heart disease

The paper by Leonard *et al*<sup>1</sup> highlights the inadequacy of contraceptive advice given to adult patients with congenital heart disease. Unfortunately, until now many patients with congenital heart lesions have been left to fend for themselves with regards to contraception and given little specific advice. The provision of insufficient or inaccurate advice has significant consequences, and consideration should be given to referring patients with complex congenital heart lesions to specific clinics where specialist gynaecological and obstetric advice can be obtained.<sup>2</sup>

### Pregnancy and the cardiovascular system

When giving family planning advice to patients with congenital heart disease it is paramount that the inherent dangers of pregnancy for that individual are weighed against the possible adverse effects of the method of contraception being considered. An understanding of the normal physiology of pregnancy underpins this assessment.

Changes in the maternal cardiovascular system are detectable even within the first two months of a normal pregnancy.<sup>3</sup> During pregnancy maternal heart rate and circulating blood volumes increase<sup>4</sup> and there is an increase in left ventricular end diastolic volume<sup>5</sup>; together these changes lead to a 30–50% augmentation of cardiac output in the normal heart.<sup>6</sup> The most dramatic swings in haemodynamic parameters occur during labour and delivery. Venous return varies greatly because of caval compression by the gravid uterus and blood loss, and the sudden contraction of an engorged uterus returning 500 ml of blood to the systemic circulation. These changes are tolerated well by the vast majority of individuals but pose a considerable challenge to the patient with congenital lesions involving poor left ventricular function, a large right to left shunt, or a morphological right ventricle in the systemic circulation—for example, those with a previous Mustard procedure.

Structural rather than functional changes are important in patients with Marfan syndrome, aortic dilatation secondary to bicuspid aortic valve disease or previous coarctation repair. In normal pregnancy serial echocardiography reveals an increase in ascending aortic dimensions<sup>7</sup> and this is of particular concern in patients already at risk of aortic dissection or aneurysm formation.<sup>8</sup>

Pregnancy is a prothrombotic state with an increase in clotting factor concentrations and platelet activity.<sup>9</sup> Thromboembolic events are more common in all women when pregnant but certain groups of patients with congenital heart disease—for example, those with Fontan repairs, poor ventricular function or atrial arrhythmias, are particularly susceptible to these events. Patients with Eisenmenger's syndrome have the additional risk of paradoxical embolic events. Many of the physiological changes associated with pregnancy persist well into the puerperium and it would be dangerous to consider the patient free from risk as soon as they deliver.

Cardiac lesions have in the past been arbitrarily, and rather simplistically, classified into "low" and "high" risk. The high risk group includes patients with New York Heart Association (NYHA) class IV symptoms, pulmonary hypertension, cyanosis, or dilated aortic roots, and those with significant left sided obstructive lesions in whom gradients worsen as systemic vascular resistance falls.<sup>10</sup> Patients with Eisenmenger's syndrome classically tolerate pregnancy poorly with a maternal mortality of up to 60%.<sup>11</sup> For patients with complex cardiac lesions an individualised pregnancy risk assessment encompassing all these elements is necessary to enable informed decision making regarding family planning.

The risks of pregnancy discussed have focused on maternal wellbeing but important and emotive issues arise regarding fetal morbidity and mortality. Insults to the developing fetus arise from an adverse intrauterine environment, with relative hypoxia and acidosis, the increased risk of congenital cardiac anomalies, and the effects of medical treatment. In addition to the teratogenic effects of oral anticoagulants, common cardiac medications such as  $\beta$  blockers and angiotensin converting enzyme inhibitors may harm the fetus resulting in an increased risk of intrauterine growth retardation<sup>12</sup> and renal tubular dysgenesis,<sup>13</sup> respectively. Invasive investigations, radiation exposure, and cardiac surgery put the fetus at risk; cardiac bypass itself is associated with a fetal loss of 20%.<sup>14</sup> Pre-pregnancy counselling and thoughtful regard to the timing of any necessary reinvestigation and intervention can simplify many of these issues.

### Contraception

Sterilisation is prescribed in a minority of patients to prevent pregnancy permanently, in others the aim of contraception is to postpone pregnancy until it suits the individual's medical and social circumstances. In an ideal world all pregnancies in this patient group would be planned allowing time for adequate pre-pregnancy counselling and assessment, ensuring that the mother-to-be is both physically and psychologically as fit as possible. Unfortunately we are far from this utopian picture but at least routine discussion of contraception in congenital heart disease practice would be a step towards this goal. When discussing contraception, patients should be assessed as individuals taking into account the nature and natural history of their cardiac lesion, concurrent medication, concomitant medical problems, thrombotic tendency, and personal wishes. One patient's perception of acceptable risk may be unacceptable to another.

### STERILISATION

Sterilisation is a highly effective method of contraception but difficult to reverse and should be considered as a permanent procedure. Unfortunately, those for whom pregnancy is most hazardous are often in the highest risk group

in terms of effective contraception.<sup>1</sup> Laparoscopic sterilisation with its associated need for anaesthesia may be problematic in the profoundly hypoxic patient and those with a history of arrhythmia. However, compared with the dangers of pregnancy sterilisation is the treatment of choice for patients with Eisenmenger's syndrome or NYHA IV symptoms. Sterilisation of the patient's partner is limited by the likelihood that the male may outlive his partner and wish to start a family with a future partner.

#### HORMONAL CONTRACEPTION

Hormonal contraception is highly effective and safe for the majority of motivated patients with simple congenital cardiac lesions. Failure rates, when used correctly, vary between 1% and 3% depending on the preparation used. There are three main preparations: the combined oral contraceptive pill, containing varying doses and forms of oestrogen and progestogen; the mini-pill (progestogen only); and the Depo preparations of progesterone (for example, Depo-provera, UpJohn).

Combined oral contraceptives have well documented adverse effects on the coagulation system<sup>15</sup> and recent controversy has surrounded the relative risks of thromboembolic events associated with varying formulations of progestogen.<sup>16</sup> It is primarily for this reason, and to a lesser extent the effects on blood pressure and lipid metabolism, that caution is exercised in prescribing these preparations to patients with cardiac lesions. Combined preparations should be avoided in those at high risk of thromboembolism or those who would tolerate thromboembolism particularly poorly. This includes patients with cyanosis, pulmonary hypertension, a low cardiac output, dilated cardiac chambers, atrial dysrhythmias, sluggish venous-conduit flow (for example in a Fontan circuit), or poorly controlled hypertension. Patients with previous thromboembolic events should avoid the combined preparations. Little is known regarding the absolute risks for these patients and many of the current recommendations are based on anecdote and commonsense. It is also unclear whether the concurrent use of anticoagulants or antiplatelet agents reduces the risk of thromboembolism. There is a clear need for a large, prospective, multicentre study of oral contraceptives in patients with congenital heart disease.

Intramuscular depot injections of medroxyprogesterone may be useful in patients with an increased risk of thromboembolism. A few cases of thromboembolism have been reported with such preparations but causality has not been established. Several subgroups of patients with congenital heart disorders have learning difficulties, including those with Down's syndrome, and compliance is a practical problem that may be overcome by parenteral contraception. These preparations are highly effective but have the disadvantage of causing menstrual irregularity and a possibility of delay in return of fertility.

Progestogen only oral preparations have a reduced incidence of thromboembolic events compared with the combined pill but require highly motivated patients as a delay in dosing as little as three hours may lead to failure. Progesterone-based contraceptives, either in oral or depot formulations, may precipitate a deterioration in ventricular function because of their fluid retaining properties, and caution should be exercised in patients with impaired systemic or pulmonary ventricular function.

#### INTRAUTERINE CONTRACEPTIVE DEVICES

Intrauterine contraceptive devices (IUCD) are a fairly effective method of contraception. They are, however, associated with an increased risk of pelvic inflammatory disease (in about 2% of users). Infection is a worry for two

reasons: the fear of endocarditis in congenital heart disease patients and the increased risk of tubal infertility. The latter has led to IUCD not usually being recommended for nulliparous women. Endocarditis associated with IUCD use is rare, probably occurring less than once per million patient-years; however, both *Streptococcus viridans* and *Neisseria gonorrhoeae* endocarditis have been reported.<sup>17</sup> IUCD should be used with caution in anticoagulated patients as a minority of individuals experience problematic uterine bleeding. These facts aside they are effective, convenient, and relatively safe for many patients. The new levonorgestrel containing IUCD may have a specific role to play in patients with menstrual bleeding problems who also require contraception.

#### BARRIER AND TRADITIONAL METHODS

Barrier and traditional methods of contraception have the advantage of having few recognised adverse effects. This does not equate with safety in the congenital group as the prime adverse effect is contraceptive failure and subsequent pregnancy. In most patients with congenital cardiac disease pregnancy is associated with far more complications than any form of contraception. For this reason all but the most motivated patients should be steered from barrier methods.

#### Conclusion

It should not be forgotten that most patients with simple congenital heart lesions—the vast majority of those attending general cardiology or family planning clinics—tolerate pregnancy and contraception well. Unfortunately those requiring more specific direction are often let down. Cardiologists are poor at raising these issues perhaps because they feel it is an area outwith their field of expertise or because they, rather protectively, perceive these patients whom they may have known since infancy, as children. Over involved parents attending outpatient clinics complicate matters. There is also a concerning tendency to state which contraceptive methods are contraindicated—for example, to advise a patient against using the combined oral contraceptive pill, without giving any positive alternatives. This inevitably leaves the patient less than protected.

When counselling patients with more complex lesions a pragmatic approach is essential. There is a finite number of contraceptive options each with their own drawbacks, and decision making may be in the form of opting for the lesser of two evils. Contraceptive advice must be tailored to the individual both in terms of their medical, educational, and social conditions. The balance between efficacy and safety is a difficult one but leaning towards efficacy is probably justified in those with more complex lesions.

Failure to give adequate contraceptive advice can have devastating consequences. Termination of pregnancy is undesirable with great potential to damage medical and psychological wellbeing. The ethical and religious convictions of the family seeking advice should be respected even in the most difficult situations. If we are confrontational patients will vote with their feet and attend only in the latter stages of pregnancy or when they run into trouble.

Despite all these difficulties, the issue of contraception should be raised with all female patients with congenital heart disease when they reach childbearing potential. Ideally this should be in the context of previous discussions, from childhood, on the lifestyle implications of the patient's cardiac disease. Although often a sensitive area, discussion about future pregnancies can be approached in much the same way as advice on career prospects or exer-

cise. Finally, it should be ensured that any method of contraception a patient with congenital heart disease uses is appropriate to their medical and social circumstances. Failure to do so constitutes a neglect of the duty of care owed to that patient and their family.

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