

# 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.<sup>1</sup> Due to recent advances in cardiac surgery and congenital heart disease, the latter is now the most common substrate of heart disease in pregnancy in the western world.<sup>2</sup> 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 intrauterine 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.<sup>3-11</sup> 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.<sup>12</sup>

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.<sup>13,14</sup> 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 al<sup>5,6</sup> and a recent meta-analysis from the Netherlands<sup>15</sup> 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.<sup>16</sup> 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).<sup>17</sup> Nevertheless, when the

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

<b>Maternal Pregnancy Related Risks (of Major Complications Including Death)</b>		
<b>Lesion Specific Risks<sup>3,4</sup></b>		
<b>Low (1:1000-1:100)</b>	<b>Moderate (1%-2%)</b>	<b>High (&gt;2%)</b>
Ventricular septal defect	Mitral stenosis	Marfan syndrome with significant aortic root/AV involvement
Atrial septal defect	Aortic stenosis	Eisenmenger syndrome
Coarctation repaired	Systemic RV (TGA after atrial switch procedure or ccTGA)	
Tetralogy of Fallot	Fontan-type circulation	Other PAH
	Cyanosis without PAH	
<b>Generic Cardiac Risks<sup>5,6,a</sup></b>		
Poor functional class before pregnancy (NYHA >II) or cyanosis		
Impaired systemic ventricular function (EF <40%)		
Left heart obstruction (MV area <2 cm <sup>2</sup> , AV area <1.5 cm <sup>2</sup> , 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		
<b>Other Risks<sup>7</sup></b>		
Age <18 and >35		
Physical deconditioning		
Smoking		
<b>Fetal Risks (of Miscarriage, IUGR, etc)<sup>5,6,8</sup></b>		
Maternal NYHA functional class >II or cyanosis		
Treatment with anticoagulants during pregnancy		
Maternal smoking		
Multiple gestation		
Left heart obstruction		
<b>Risk of Transmission of CHD,<sup>4,b</sup> %</b>		
	<b>Mother Affected</b>	<b>Father Affected</b>
Atrioventricular septal defect	11.6	4.3
Aortic stenosis	8.0	3.8
Coarctation	6.3	3.0
Atrial septal defect	6.0	3.6
Pulmonary stenosis	5.3	3.5
Tetralogy of Fallot	2.0	1.4
<b>Maternal and Paternal Survival Prospects<sup>19</sup></b>		
Should be discussed:	Parental life expectancy	
	Risk of premature death or disability	
	Need for major surgery	
<b>Levels and Intensity of Health Care Required<sup>3</sup></b>		
<b>Pregnancy</b>		
<b>Low Risk Patients</b>	<b>Moderate and High Risk Patients</b>	
Local hospital	Tertiary hospital	
1 visit at ACHD clinic (clarify issues/make recommendations)	Multidisciplinary approach "Joint Cardiac Obstetric Clinic"	
<b>Delivery and Postpartum</b>		
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)		

(Continued)

## Preconception Counselling for Women With Congenital Heart Disease (Continued)

### Effective Contraception and Termination of Pregnancy<sup>10</sup>

#### 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

ACHD, indicates adult congenital heart disease; AV, aortic valve; ccTGA, congenitally corrected TGA; CHD, congenital heart disease; IUGR, intrauterine growth restriction; LV, left ventricle; NYHA, New York Heart Association functional classification; MV, mitral valve; PAH, pulmonar arterial hypertension; RV, right ventricle; TGA, transposition of the great arteries;

<sup>a</sup>Expected cardiac event rate in pregnancies with 0, 1, or >1 of these factors is 5%, 27%, and 75%, respectively.

<sup>b</sup>General population risk, 0.8%; autosomal dominant heritability [DiGeorge (22q11), Marfan, Noonan's syndromes], 50%.

individual patient decides to proceed with pregnancy or presents pregnant to us, all efforts should be made to minimize risks involved. To achieve this objective a multidisciplinary approach, additional resource allocation, and great attention to detail are all essential.<sup>8,18</sup>

Manso et al<sup>19</sup> in this issue of the journal report their experience on pregnancy outcomes amongst 56 women with congenital heart disease. Severity of congenital heart disease had an impact on both maternal and neonatal outcomes. The authors should be congratulated for their multidisciplinary approach towards this new challenge and for the opportunities that derive both for health care professionals as well as patients with congenital heart disease. Congenital heart disease is the most common congenital defect (approximately 1% of newborns are affected around the world). At least 85% of patients with congenital heart disease now survive to adulthood and half of them are women, most of reproductive age. Their natural desire is to have their own children. We, cardiologists, clearly need to work closer with our obstetric and anaesthetic colleagues, understand better the impact of pregnancy on long-term cardiovascular outcome, and communicate at greater extent this emerging data with health care professionals and our patients, so that we can help every single patient with congenital heart disease reach their full life potential.

### REFERENCES

1. Saving Mothers' Lives: Reviewing maternal deaths to make motherhood safer — 2003-2005. The seventh report of the Confidential Enquiries into Maternal Deaths in the United Kingdom. London: RCOG Press; 2007. Available from: [www.cemach.org.uk](http://www.cemach.org.uk)
2. Wren C, O'Sullivan JJ. Survival with congenital heart disease and need for follow up in adult life. *Heart*. 2001;85:438-43.
3. Steer PJ, Gatzoulis MA, Baker PN. Heart disease and pregnancy. London: RCOG Press; 2006.
4. Uebing A, Steer PJ, Yentis SM, Gatzoulis MA. Pregnancy and congenital heart disease. *BMJ*. 2006;332:401-6.
5. Siu SC, Sermer M, Harrison DA, Grigoriadis E, Liu G, Sorensen S, et al. Risk and predictors for pregnancy-related complications in women with heart disease. *Circulation*. 1997;96:2789-94.
6. Siu SC, Sermer M, Colman JM, Alvarez AN, Mercier LA, Morton BC, et al. Cardiac Disease in Pregnancy (CARPREG) Investigators. Prospective multicenter study of pregnancy outcome in women with heart disease. *Circulation*. 2001;104:515-21.
7. Steer PJ. Pregnancy and contraception. In: Gatzoulis MA, Swan L, Therrien J, Pantely GA, editors. Adult congenital heart disease: a practical guide. Oxford: BMJ, Blackwell; 2005. p. 16-35.
8. Khairy P, Ouyang DW, Fernandes SM, Lee-Parritz A, Economy KE, Landzberg MJ. Pregnancy outcomes in women with congenital heart disease. *Circulation*. 2006;113:517-24.
9. Nieminen HP, Jokinen EV, Sairanen HI. Late results of pediatric cardiac surgery in Finland: a population-based study with 96% follow-up. *Circulation*. 2001;104:570-5.
10. Dhanjal MK. Contraception in women with heart disease. In: Steer PJ, Gatzoulis MA, Baker P, editors. Heart Disease and Pregnancy. London: RCOG; 2006. p. 9-26.
11. Hunter S, Robson SC. Adaptation of the maternal heart in pregnancy. *Br Heart J*. 1992;68:540-3.
12. Niwa K, Tateno S, Akagi T, Himeno W, Kawasoe Y, Tatebe S, et al. Arrhythmia and reduced heart rate variability during pregnancy in women with congenital heart disease and previous reparative surgery. *Int J Cardiol*. 2007;122:143-8.
13. Weiss BM, Zemp L, Seifert B, Hess OM. Outcome of pulmonary vascular disease in pregnancy: a systematic overview from 1978 though 1996. *J Am Coll Cardiol*. 1998;31:1650-7.
14. Presbitero P, Somerville J, Stone S, Aruta E, Spiegelhalter D, Rabajoli F. Pregnancy in cyanotic congenital heart disease. Outcome of mother and fetus. *Circulation*. 1994;84:2673-6.
15. Drenthen W, Pieper PG, Roos-Hesselink JW, van Lottum WA, Voors AA, Mulder BJ, et al. Outcome of pregnancy in women with congenital heart disease: a literature review. *J Am Coll Cardiol*. 2007;49:2303-11.
16. Guedes A, Mercier LA, Leduc L, Berube L, Marcotte F, Dore A. Impact of pregnancy on the systemic right ventricle after a Mustard operation for transposition of the great arteries. *J Am Coll Cardiol*. 2004;44:433-7.

17. Gelson E, Gatzoulis M, Johnson M. Valvular heart disease. *BMJ*. 2007;335:1042-5.
18. Task Force on the Management of Cardiovascular Diseases during Pregnancy of the European Society of Cardiology. Expert consensus document on management of cardiovascular diseases during pregnancy. *Eur Heart J*. 2003;24:761-81.
19. Manso B, Gran F, Pijuan A, Giral G, Ferrer Q, Betrián P, et al. Embarazo y cardiopatías congénitas. *Rev Esp Cardiol*. 2008; 61:236-43.