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REVIEW

Clinical update

Pregnancy in women with congenital heart disease

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Congenital heart defects are the most common birth defects. Major advances in open-heart surgery have led to rapidly evolving cohorts of adult survivors and the majority of affected women now survive to childbearing age. The risk of cardiovascular complications during pregnancy and peripartum depends on the type of the underlying defect, the extent and severity of residual haemodynamic lesions and comorbidities. Careful individualized, multi-disciplinary pre-pregnancy risk assessment and counselling, including assessment of risks in the offspring and estimation on long-term outcomes of the underlying heart defect, will enable informed decision making. Depending on the estimated risks, a careful follow-up plan during pregnancy as well as a detailed plan for delivery and postpartum care can reduce the risks and should be made by the multi-disciplinary team.

Keywords Pregnancy • Congenital heart disease • Labour • Delivery

Background and scope of the problem

The field of pregnancy in women with heart disease is dynamic and novel evidence is constantly evolving. The goal of this article is to give an overview on how to incorporate this growing knowledge into day-to-day practice regarding counselling, decision making, and management of women with congenital heart disease before, during, and after pregnancy.

Congenital heart defects (CHDs) affect 0.4–1.5% of the general population.[1–3] Nowadays due to improved surgical and medical treatment, survival to adulthood has become the rule even for patients with the most complex lesions.[4] This has resulted in a large cohort of adult survivors, mostly still young and including many women in childbearing age.[5,6] It is important to recognize that these patients are not cured and many are at high risk of cardiovascular complications and increased risk of premature death.[6]

Pregnancy-related cardiac deaths have increased over the last two decades.[7] The proportion of pregnancy-related deaths attributed to CHD has, however, remained relatively low (see *Figure 1*). This is in spite that in most clinics specialized in heart disease in pregnancy in developed countries; women with CHD comprise the largest group of patients.[8,9] This may be partly explained by the fact that most women with CHD are followed at specialized centres and thus receive appropriate pre-conception assessment and counselling, as well as structured follow-up during pregnancy. In contrast, women with acquired heart disease sometimes present the first time during pregnancy with a complication, such as myocardial infarction, arrhythmia, or aortic dissection. The prevalence of ischaemic heart disease complicating pregnancies is increasing, perhaps related to trends in Western societies of having children later in life and associated higher rates of cardiovascular risk factors in these older women.[7,10,11]

Given the epidemiology of CHD with rapidly expanding cohorts of young women with complex CHD (i.e. Fontan palliation for univentricular hearts), we have to prepare for an increasing number of high-risk pregnancies.

Individual pregnancy risks: enabling informed decision making

Haemodynamic changes during pregnancy, delivery, and postpartum

Pregnancy and delivery require profound adaptations of the cardiovascular system to cope with the increased haemodynamic needs.

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The extent of these haemodynamic changes in healthy women is illustrated in *Figure 2*.

Depending on the type and complexity of the congenital cardiac defect and the severity of residual haemodynamic lesions, these normal adaptive responses of the cardiovascular system to the increased demands of pregnancy, labour, and puerpurium can be distorted in numerous ways.[16] The occurrence of tachy-arrhythmias or failure to adequately increase heart rate or cardiac output (chronotropic incompetence, inadequate adaptation of pacemaker programming, valve stenosis, rigid conduits, or baffles) during pregnancy and peripartum may further impair these adaptive mechanisms and precipitate decompensation.

The occurrence of obstetric complications, such as hypertensive disorders of pregnancy, pre-eclampsia, or multiple pregnancies increase the haemodynamic load of pregnancy and thus increase the risk of maladaptation to haemodynamic needs and subsequently increase the risk of cardiovascular complications in vulnerable patients.[8,17–19] The increased risk of multiple pregnancies may be of particular importance in women seeking assisted fertilization and underscores the need for optimal multi-disciplinary pre-pregnancy communication, counselling, and planning. Assisted reproductive technologies carry in it risks that may endanger the pregnant woman with cardiovascular disease, such as fluid retention and enhancement of hypercoagulability.[20]

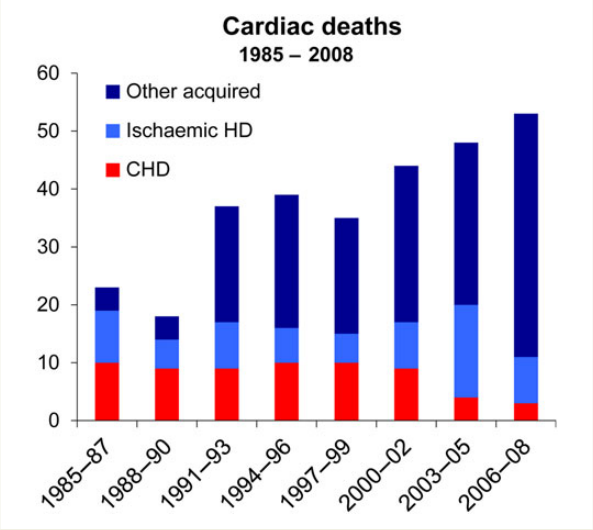

Figure 1 Number of maternal deaths related to cardiac disease in the UK between 1985 and 2008, stratified for underlying heart condition (modified from Cantwell *et al.*[7]).

Risk assessment, counselling, and decision making

Risk stratification in CHD is complicated by the diversity of lesions and surgical repair techniques as well as by the variety of (residual) haemodynamic lesions.

Guidelines recommend that all women with CHD should have careful pre-pregnancy assessment and detailed pre-pregnancy counselling prior to embarking on pregnancy. To fulfil this task, a comprehensive assessment by experienced cardiologists, obstetricians, and specialists from other specialities is needed. Since most women with CHD are followed at specialized centres, the opportunities for such counselling are generally available. Nonetheless, studies show that in reality counselling may be suboptimal.[21–24]

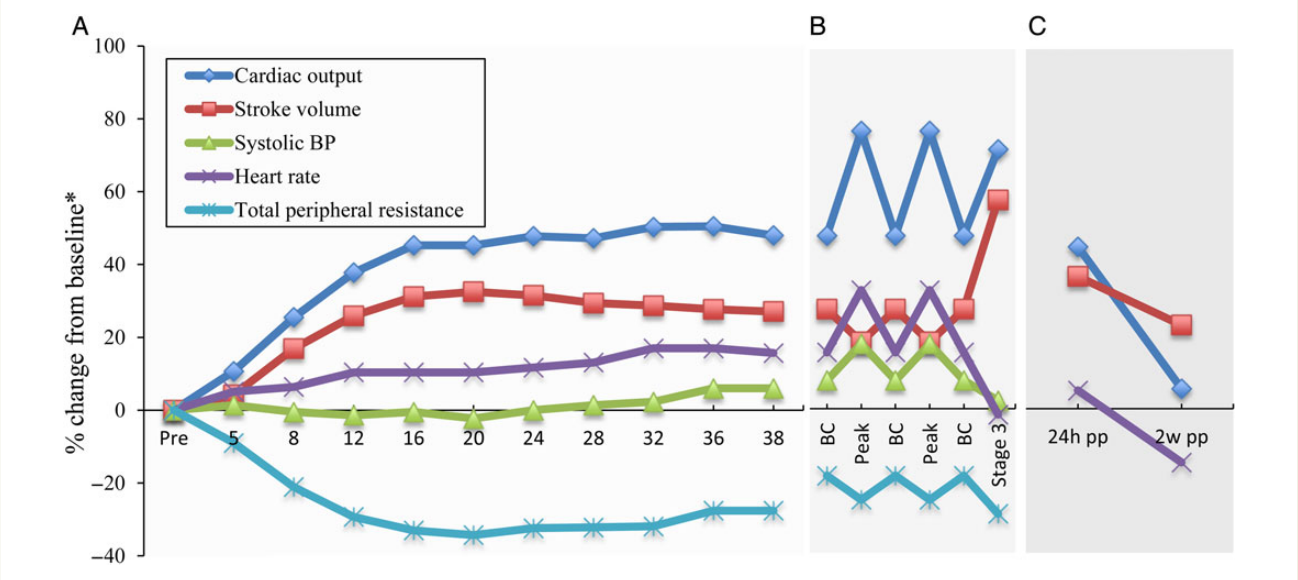

Figure 2 Haemodynamic changes during pregnancy, peripartum, and postpartum. (*A*) Pregnancy (weeks of gestation). Modified from Robson *et al.*[12] (*B*) Peripartum. Modified from: Adams *et al.*[13] BC, between contractions; Peak, at the peak of contraction; Stage 3, at the time of uterine contraction. (*C*) Postportum. Modified from Robson *et al.*[14,15] 24 h pp, 24 h postpartum; 2w pp, 2 weeks postpartum. *For cohorts in (*B*) and (*C*), relative changes from baseline were compared with the baseline values of the cohort from (*A*).

Over the last few decades, there has been rapidly growing evidence on pregnancy risks for a variety of CHD. One source of evidence stems from lesion-specific studies. Most of these studies are retrospective studies and often comprise relatively small numbers of pregnancies at risk and a wide variety of residual haemodynamic lesions within specific groups of CHD. With the advent of large multi-centre registries, such as the ROPAC registry, more detailed data on lesion-specific outcomes will help better individual risk stratification in the future. A meta-analysis summarizing the results of lesion-specific studies, published in 2007 by the ZAHARA investigators, provides a useful estimation of the spectrum of risks for a given CHD.[25] All these studies are however flawed by the important biases of such small retrospective series and therefore must be used carefully in individual decision making. Another source of evidence stems from studies on larger numbers of women with various forms of heart diseases and thus could define risk factors for adverse outcomes based on patient history and residual haemodynamic lesions and define risk scores.[8,9,16] Based on these sources of evidence and on collective clinical experience, expert panels have defined risk-categories based on the underlying CHD, specific haemodynamic residue as well as past medical history (see *Figure 3* and corresponding *Box 1*). This so-called modified WHO classification of maternal pregnancy risk has prospectively been validated as the most reliable method of risk assessment.[26] Though this is a helpful tool for risk estimation, it should be emphasized that risk stratification must be individualized (see *Figure 3*).

The assessment of risk for cardiovascular and foetal complications in women with CHD is a *multi-disciplinary* (see *Figure 4*) and *multi-step* process, beginning with a detailed exploration of the patient's previous history, electrocardiogram, detailed echocardiography and additional testing, such as exercise testing or advanced imaging, as appropriate. This process is outlined in *Figure 3* and the associated boxes. For individual decision making, it is very important not only to estimate the probability of any adverse outcomes but to distinguish between complications that can usually be well managed (i.e. mild heart failure, atrial arrhythmias) and complications that may be fatal or lead to permanent damage or deterioration (i.e. stroke, irreversible deterioration of ventricular function).

An important aspect of risk stratification is to decide, whether a pre-pregnancy intervention, be it medication or an invasive procedure, may reduce the risk. This requires thought-out decision making since sometimes the 'cure is worse than the disease'. For example, the risk of serious adverse cardiac events may be higher in women with a mechanical aortic valve prosthesis compared with an asymptomatic woman with severe aortic stenosis and normal haemodynamic response to exercise. Furthermore, premature interventions, such as prophylactic pulmonary valve replacement in women with pulmonary regurgitation may expose the woman to an uncertain long-term risk because of the life-time need for multiple subsequent interventions and higher risk of endocarditis, while there is no proven evidence for risk reduction during pregnancy and long term.[36,37]

Pre-pregnancy assessment must include a careful review of all medications. Some cardiac medications, such as angiotensin-converting-enzyme inhibitors, angiotensin- or aldosterone-antagonists, are contraindicated during pregnancy. In women taking such medications, careful estimation of risk of being off such medications for a certain time is mandatory. It may be wise to test the effect of cessation of such medications on ventricular function and well-being prior to embark on pregnancy.

Box 1 Factors associated with high maternal risk[8,18,22,25,27]

Congenital heart defects/residual hemodynamic lesions with very high maternal risk

(High risk of maternal mortality, consensus that pregnancy should be discouraged)

- Severe symptomatic aortic valve stenosis
- Severe left atrio-ventricular valve stenosis
- Pulmonary arterial hypertension
- Systemic ventricular ejection fraction <30%
- Poor functional class (NYHA III and IV)
- Marfan syndrome with ascending aortic diameter >45 mm
- Bicuspid aortic valve with ascending aortic diameter >50 mm
- Unrepaired severe coarctation

Congenital heart defects/residual haemodynamic lesions with high maternal risk[8,18,22,25,27]

(High risk of morbidity or increased risk of maternal mortality, consensus that very careful pre-pregnancy assessment and counselling is required)

- Mechanical heart valve prosthesis
- Systemic right ventricle (transposition of the great arteries after atrial switch operations or congenitally corrected transposition of the great arteries)
- Fontan palliation for univentricular hearts
- Unrepaired or palliated cyanotic heart conditions
- Marfan syndrome with ascending aortic diameter <40 mm
- Severe systemic atrio-ventricular valve regurgitation
- Asymptomatic left ventricular outflow tract stenosis with peak gradient >50 mmHg or aortic valve area <1.5 cm^2
- Left atrio-ventricular valve stenosis with valve area opening <2.0 cm^2
- Systemic ventricular ejection fraction 30–40%

Foetal risk

In many women with congenital heart disease, the risk of miscarriage is substantially increased. Foetal risks comprise mainly the risk of prematurity, low birth weight/small for gestational age, offspring mortality, and the risk of recurrence of CHD. The risk of prematurity and low birth weight/small for gestational age is closely related to the severity of maternal heart disease and may be extremely high in women with the most severe forms, such as univentricular physiology, Eisenmenger-physiology, and unrepaired cyanotic defects. Prematurity can result from spontaneous premature labour but also from early-induced delivery for maternal (cardiac) reasons.[25] Individual risk factors that have been associated with adverse foetal outcomes are listed in *Box 2*. However, risk scores developed from these risk factors did not perform well in predicting foetal complications in a recent prospective study.[26] The association of complexity of maternal heart disease and foetal outcome underscores the importance of our multi-disciplinary efforts to maintain optimal maternal haemodynamics and well-being during pregnancy. Foetal outcome is associated with uteroplacental blood flow. Recent evidence suggests that uteroplacental blood flow is associated with maternal haemodynamics in women with CHD. Impaired uteroplacental

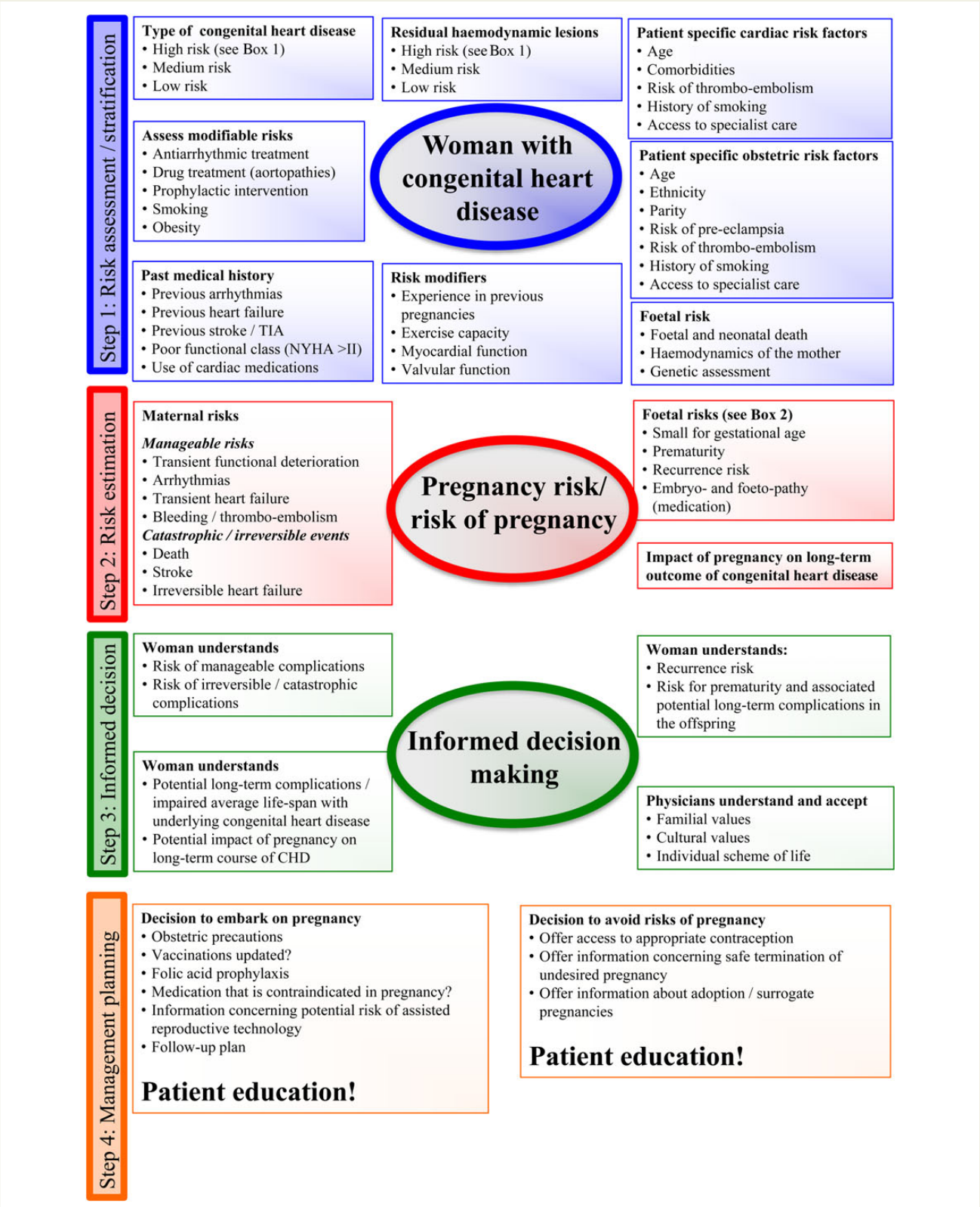

Figure 3 Multi-disciplinary, multi-step risk assessment, counselling, and management planning (Refs 6,8,18,22,25,27–34).

blood flow may be the result of maternal impaired haemodynamics, explaining partly the increased foetal complication rate in women with CHD.[38]

The recurrence risk of isolated CHDs in the offspring is on average ~3–5%.[39] There is however substantial variability in recurrence risk, depending on familial occurrence and type of defect.[40,41] The

recurrence risk is particularly high in left ventricular outflow tract obstruction, including the hypoplastic left heart syndrome.[42] Some CHDs are associated with syndromes such as the 22q11 microdeletion syndrome (cono-truncal defects), Noonan syndrome (pulmonary stenosis, atrial septal defects), and the Holt-Oram-syndrome (atrial and ventricular septal defects). In these syndromes, inheritance is autosomal dominant and recurrence risk is 50%. Genetic testing using cord blood samples can be discussed with the affected couple. In all patients with CHDs, including male patients, recurrence risk should be discussed and counselling by a geneticist should be offered. Counselling by a geneticist may be particularly important in patients asking for pre-implantation testing in the context of assisted fertilization.

Effect of pregnancy on long-term outcomes

Only few studies have assessed the impact of pregnancy on long-term outcomes in women with heart disease. For women with subaortic right ventricles in the setting of transposition of the great arteries after atrial switch operations, there is a small but definitive—and unpredictable—risk of irreversible deterioration of systolic dysfunction of the subaortic systemic right ventricle.[28,43,44] A case–control study in women with dilated cardiomyopathy and systemic ventricular dysfunction suggests an increased short-term risk of adverse cardiovascular events in women who went through a pregnancy compared with matched controls without pregnancies.[29] Similarly, women with aortic stenosis who went through pregnancy have more events than non-pregnant controls.[45] Several studies have shown that women with CHD who have an adverse event during pregnancy are more likely to experience adverse events after pregnancy than women with uncomplicated pregnancies.[46,47]

A discussion about these uncertainties about the long-term effect of pregnancy on outcomes of the underlying heart defect may thus be offered to the women with CHD.

Prognosis of congenital heart condition

The prognosis of the woman with CHD may interfere with her ability to raise children. This should be discussed when applicable, even though[34,48] such discussions are difficult as prognostication may be uncertain and a number of barriers impair these discussions.[48] The discussion about pregnancy risks may however be a good setting to explore the women's wishes to discuss such issues. It is well known that patient knowledge about long-term risks of the underlying heart defects is often inappropriate.[49] To detain discussions about long-term prognosis of the underlying CHD may impede informed decision making about family planning in affected women. It should also be emphasized that it is important to offer these discussions not only to our female patients but also to our male patients when they contemplate to start a family!

Management planning

Once a woman has made a decision whether or not to accept the risks of a pregnancy, it is important to plan future management. For women not wishing to become pregnant, safe and effective contraception should be provided.[27,32] When there is no current but possible future pregnancy wish, the woman should be instructed to use safe and effective contraception and when she desires to embark on a pregnancy, to visit her cardiologist for an up-to-date assessment of pregnancy risk (including echocardiography and exercise testing) and construction of a pregnancy management plan before discontinuing her contraceptive.

For women, who wish to become pregnant it is important to clearly define management during pregnancy and to provide obstetric assessment to ascertain up-to-date vaccination and initiation of appropriate prophylactic measures, such as folic acid supplementation as well as recommendations for smoking cessation and abstinence from alcohol.

Management of pregnancy in women with congenital heart disease

General aspects

For each pregnancy, a clear follow-up plan must be provided. The content of this plan is dependant on the estimated maternal and foetal risk. While high-risk pregnancies (see *Figure 3*, *Box 1*) must be managed by a dedicated, experienced multi-disciplinary team at a tertiary care centre, women at low risk for cardiovascular and foetal complications can often be managed at regional centres. Good collaboration between regional centres and tertiary referral centres is of paramount importance. All pregnant women with CHD should be offered foetal echocardiography between 18 and 21 weeks of gestation.

The frequency of cardiologic follow-up visits during pregnancy must be individualized according to estimated risks and anticipated complications based on the comprehensive risk assessment (see *Figure 3*). For women with low or intermediate risk, assessment towards the end of the first trimester, ~20 weeks of gestation and at peak of haemodynamic load at ~28–32 weeks of gestation is appropriate. Those at high risk need monthly or bimonthly follow-up.[22] Dependent on individual risk stratification and the course during pregnancy, follow-up may however significantly differ from this scheme. The importance of patient education cannot be overemphasized. Every woman, who experiences novel or worsening symptoms during pregnancy (i.e. palpitations or increasing shortness of breath) should be encouraged to seek prompt assessment by her specialized multi-disciplinary team.

The woman who had not had pre-conception assessment and counselling

Occasionally, women present first to a specialist multi-disciplinary centre when already pregnant. In these patients, assessment and risk stratification follows generally the same multi-disciplinary, stepwise approach as pre-conception counselling and assessment, although with certain variations.

Basis of the assessment is a comprehensive evaluation of the patient's history, a thorough physical examination, an electrocardiogram, and a detailed echocardiography. However, as a comprehensive assessment of maternal health is of paramount importance for the estimation of maternal and foetal risk of adverse events, additional testing may be required.

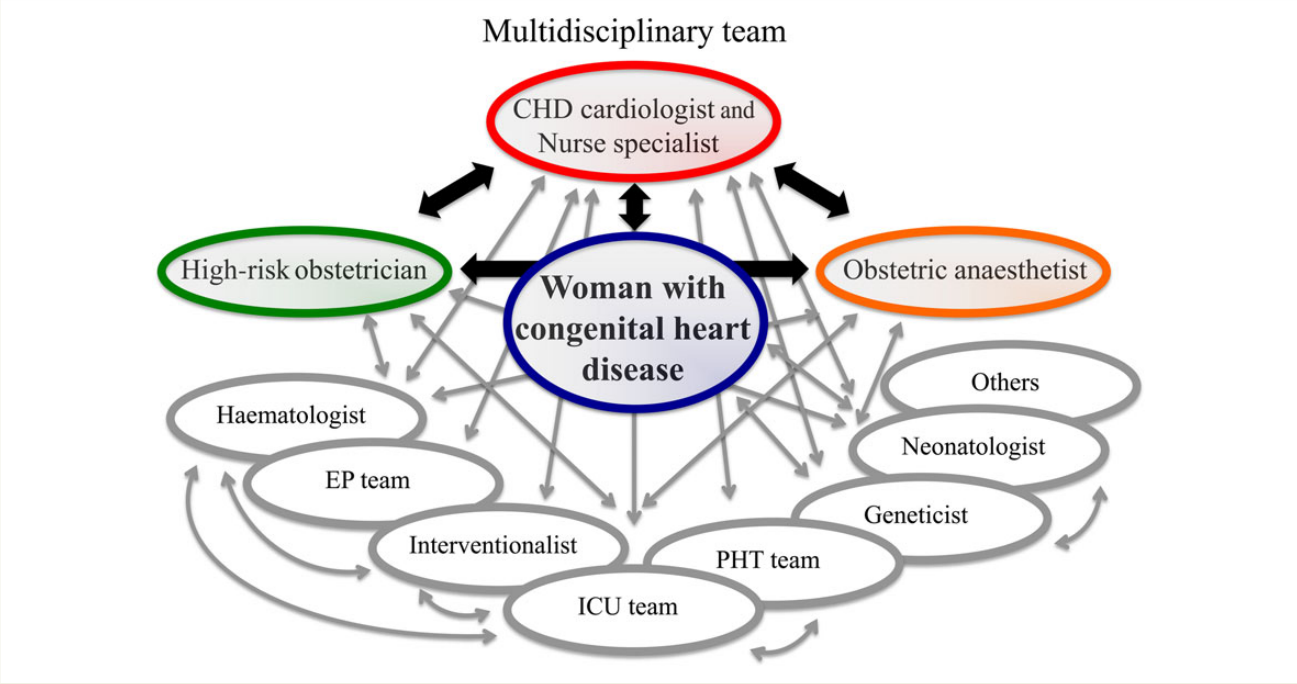

Figure 4 Multi-disciplinary team. CHD, congenital heart disease; EP, electrophysiology; ICU, intensive care unit; PHT, pulmonary hypertension. Additional teams and services may need to be involved (i.e. social service) as per individual needs of the pregnant woman. In some services, maternal foetal medicine specialists are part of the team.

Box 2 Risk of neonatal events[8,18,35]

Maternal pre-pregnancy functional class >II or cyanosis
Maternal left heart obstruction
Smoking during pregnancy
Multiple gestation
Use of oral anti-coagulants during pregnancy
Mechanical heart valve prosthesis

Submaximal exercise testing to evaluate exercise capacity, heart rate, and blood pressure response to exercise and exercise-induced arrhythmias can be safely performed during pregnancy.[22] Given the high chance of spontaneous miscarriage before 12 weeks, it may however be prudent to avoid exercise testing in the first trimester, if possible, as any temporal relationship between miscarriage and exercise testing may be misconstrued as causal rather than pure chance. When an exercise test is performed, the aim is to achieve 80% of the predicted heart rate. As chronotropic incompetence is common in adults with CHD, cardio-pulmonary exercise testing with measuring gas-exchange, aiming for a respiratory exchange ratio of 1.0 may be an alternative to standard bicycle or treadmill exercise testing.

The use of cardiac magnetic resonance imaging is regarded safe, particularly after the first trimester, but the use of gadolinium-contrast should be avoided. This may be particularly indicated for imaging of the entire aorta in women with Marfan syndrome or other aortic diseases. Methods using ionizing radiation (radiography, computed tomography, or scintigraphy) are generally avoided, unless they provide critical information that cannot be obtained with other methods.[22]

If the assessment reveals a very high-risk condition (see *Figure 3*, *Box 1*), termination of pregnancy should be discussed, preferably early in pregnancy. However, termination of pregnancy carries risks and for women with high-risk lesions this should be performed in a tertiary centre.

Need for intervention or cardiac surgery during pregnancy

Although percutaneous interventions are generally possible during pregnancy, the decision to perform such an intervention requires careful interdisciplinary assessment with weighing risks and benefits. Procedures should be performed by experienced teams at tertiary care centres only, with an aim to keep the radiation dose to the foetus as minimal as possible by shortening radiation time, using brachial artery access and shielding of the uterus. If possible, the intervention should be performed in the second trimester after completion of organogenesis but before the uterus has become very large. The timing of interventions remains debateable, however. Open-heart surgery on cardio-pulmonary bypass carries a high risk of foetal loss. If cardiac surgery can be delayed until the 28th week of gestation, prior delivery after lung maturity induction should be considered.[22]

Occurrence and management of heart failure in pregnancy and peripartum

A recent multi-centre study has shown two peaks for the occurrence of heart failure in women with heart disease.[50] One peak was towards the end of the second trimester of pregnancy, when

the cardiovascular demands reach their plateau and the second peak was peri- and early postpartum. It is however important to emphasize that the manifestation of heart failure may differ between different types of CHD as well as between individuals.

Towards later stages of pregnancy diagnosis of heart failure may be difficult as its signs and symptoms (shortness of breath on exertion, peripheral oedema, and sinus tachycardia) may be difficult to distinguish from normal findings in late pregnancy. Clinical judgment of fluid status is important and must be supplemented by regular laboratory and echocardiographic assessments. A chest X-ray is relatively harmless for the foetus and can be performed when required. Serial measurements of natriuretic peptides (Pro-BNP or BNP) may help in risk stratification.[30,33] Normal levels of natriuretic peptides at 20 weeks of pregnancy seem to provide a good negative predictive value regarding the occurrence of cardiovascular complications. Although elevated or rising levels of natriuretic peptides do not necessarily predict cardiovascular complications, it may be prudent to follow these women closely.

If overt heart failure occurs during pregnancy, patients should be admitted to a tertiary care centre. Precipitating factors, such as (intermittent) arrhythmia, should be actively sought and excluded. Bed rest, supplemental oxygen, and careful fluid balance with daily weight measurements should be initiated. Inotropes may be needed to improve heart failure and careful administration of diuretics may be used to improve pulmonary congestion but overdiuresis with worsening uteroplacental blood flow must be avoided.[51] Apart from β-blockers, standard heart failure treatment such as angiotensin-converting-enzyme inhibitors or aldosterone-antagonists is contraindicated in pregnancy. Hydralazine and nitrates may be used for afterload reduction.[22] After delivery, the use of angiotensin-converting-enzyme inhibitors is usually safe for the baby in breast-feeding women. A detailed list regarding recommendations for drug use during pregnancy and lactation can be found within the ESC guidelines.[22] Good thromboembolic prophylaxis is mandatory, particularly in patients with bed rest. In the case of refractory heart failure delivery should be contemplated as soon as the foetus is viable, or, in the case of persistent haemodynamic instability, irrespective of the duration of gestation, as maternal health always has priority. Lung maturity induction with corticosteroids may lead to fluid retention and worsening of heart failure. After restoration of haemodynamic stability postpartum, afterload reduction with angiotensin-converting-enzyme inhibitors should be started.

In women with cardiac pacemakers and implanted cardiac defibrillators it is important to document the current programming of the device (i.e. the heart rate, when anti-tachycardia pacing or defibrillation will be administered). In the case of any uncertainties, it is advisable to consult the patient's cardiologist and/or electrophysiologist in order to discuss the need for modifications of pacemaker or defibrillator programming.

Women with mechanical heart valves

Women with mechanical heart valves are at high risk of thromboembolic events.[52–54] Several strategies of anti-coagulation have been reported.[53,55,56]Anti-coagulation with vitamin K antagonists throughout pregnancy is safest for the mother and provides the lowest risk for valve thrombosis.[22,53] Offspring of women requiring low doses of vitamin K antagonists (<5 mg Warfarin or <3 mg phenprocoumon) to maintain therapeutic international normalized ratios have a low risk of embryopathy and foetal loss compared with those requiring higher doses.[52,57–61] Continuation of the vitamin K antagonist throughout pregnancy should be considered in these women.[22] Heparins are associated with more valve thrombosis but do not cross the placenta and thus do not carry the risk of embryopathy. Subplacental bleeding with the risk of pregnancy loss is associated with all anti-coagulation regimens.[52–54,58] Importantly, anti-coagulation with low-molecular-weight heparin without meticulous monitoring of anti-factor Xa levels is inappropriate and dangerous.[62] All pregnant women with mechanical heart valves need to be closely followed by a multi-disciplinary team at a tertiary care centre with a high-risk pregnancy program.[22] A detailed management plan regarding the anti-coagulation regimen during delivery and peripartum is particularly important.

Arrhythmias

Arrhythmias are by far the most common complication in adults with CHD and complicate a substantial number of pregnancies.[8,9,18,63,64] The most common type of arrhythmia is atypical atrial flutter (intra-atrial re-entrant tachycardia) caused by atrial scars.

Table 1 Obstetric drugs, their cardiovascular side effects and precautions in women with congenital heart disease

	Cardiovascular side effects	Contraindications and precautions
Drugs to treat premature contractions		
β-Mimetics (i.e. hexoprenaline)	Tachycardia, arrhythmias	Obstruction of left-sided atrio-ventricular valve Propensity for arrhythmias
Atosiban (oxytocin antagonist)	None	None
Drugs for induction of labour		
Misoprostol	Coronary vasospasm, arrhythmias	
Oxytocin	Systemic hypotension	Only low-dose continuous infusion or small repeated bolus
Drugs to control postpartum bleeding		
Oxytocin	Systemic hypotension	Only low-dose continuous infusion or small repeated bolus
Prostaglandin F analogues	Increase in pulmonary pressures	Pulmonary hypertension, right ventricular failure

Arrhythmias increase the risk of thromboembolic complications and are an important trigger for the occurrence of heart failure. Women with new-onset palpitations or sudden worsening of shortness of breath thus require prompt assessment by the multidisciplinary team. Most women with sustained or severely symptomatic arrhythmias during pregnancy need to be admitted. Depending on the underlying cardiac condition and haemodynamic stability a rate- or rhythm control strategy must be chosen, although rhythm control is often preferred over rate control in pregnant women with CHD and new-onset arrhythmias. Regarding antiarrhythmic drug treatment, β-blockers are first choice for most arrhythmias. A detailed list regarding recommendations for drug use during pregnancy and lactation can be found within the ESC guidelines.[22] Careful assessment for the need for anti-coagulation is mandatory.

Box 3 Conditions in which caesarean section is preferred over vaginal delivery

- Women with an ascending aorta diameter >45 mm
- Pre-term labour while on oral anti-coagulation
- Women with severe aortic stenosis experiencing symptoms during pregnancy (preferably general anaesthesia and endotracheal intubation)
- Severe heart failure

Obstetric complications and their impact on cardiac disease

Pre-eclampsia and multiple pregnancies may be important modifiers of the risk of cardiovascular complications in women with CHD. In the ROPAC registry (a prospective multi-centre, multi-national registry led by the European Society of Cardiology), women with structural heart disease who developed pre-eclampsia had a 30% risk of heart failure.[50] Women at risk for pre-eclampsia should be

Box 4 Requirements for a comprehensive multi-disciplinary delivery plan

1. **Information on the underlying heart defect and current haemodynamics**
   - ☑ Exact cardiac diagnosis, previous surgical, and interventional procedures
   - ☑ In the case of complex cardiac or venous anatomy, a picture of the woman's heart is advisable
   - ☑ Previous cardiovascular complications
   - ☑ Current cardiovascular findings and important residual lesions
2. **Specific recommendations and pitfalls**
   - ☑ Use of air bubble filters (see *Figure 5*) in women with risk of paradoxical embolism
   - ☑ Site of blood pressure measurements in women with occlusion of arm arteries (i.e. after subclavian flap repair for coarctation of the aorta or previous classical Blalock-Taussig-Shunts)
3. **Anticipated complications**
   - ☑ Type and expected timing of the occurrence (i.e. pulmonary oedema in Stage 3 of delivery in women with left-sided atrio-ventricular valve obstruction)
   - ☑ Suggested management in the case of complications (i.e. arrhythmias, heart failure)
   - ☑ Contact details of involved cardiologists in case of complications
   - ☑ List of specific drugs that need to be immediately available on the labour ward. In the case of drugs that are not commonly used on the labour ward (i.e. amiodarone) clear instructions on how to prepare and use such drugs is mandatory.
4. **Caution about obstetric drugs if adverse effects are expected (see *Table 1*)**
   - ☑ β-Mimetics (i.e. hexoprenaline): ☐ no restriction ☐ caution ☐ do not use
   - ☑ Atosiban (oxytocin antagonist): ☐ no restriction ☐ caution ☐ do not use
   - ☑ Misoprostol: ☐ no restriction ☐ caution ☐ do not use
   - ☑ Oxytocin: ☐ no restriction ☐ caution ☐ do not use
   - ☑ Prostaglandin F analogues: ☐ no restriction ☐ caution ☐ do not use
5. **Detailed list of medications**
6. **Allergies**
7. **Information on important obstetric issues**
   - ☑ Previous obstetric history and complications
   - ☑ Estimated date of delivery
8. **Recommendation for delivery**
   - ☑ Mode and timing of delivery (i.e. spontaneous delivery or induction at pre-specified date)
   - ☑ Recommendation for anaesthesia during delivery
   - ☑ Recommendation for rhythm monitoring during labour and delivery
   - ☑ Recommendation for type and duration of haemodynamic monitoring during labour and delivery
9. **Detailed plan for postpartum care**
   - ☑ Place of postpartum care (intensive care unit, cardiology ward, and obstetric ward)
   - ☑ Necessity and duration of rhythm monitoring postpartum
   - ☑ If required, recommendation for type and duration of haemodynamic monitoring postpartum
   - ☑ Recommendation for duration of postpartum stay in hospital (24 h/48 h/72 h)
   - ☑ Specific precautions and treatment recommendations (i.e. careful fluid balance)
   - ☑ Recommendation for investigations postpartum (i.e. pre-discharge echocardiography)
10. **Follow-up plan after discharge from hospital**

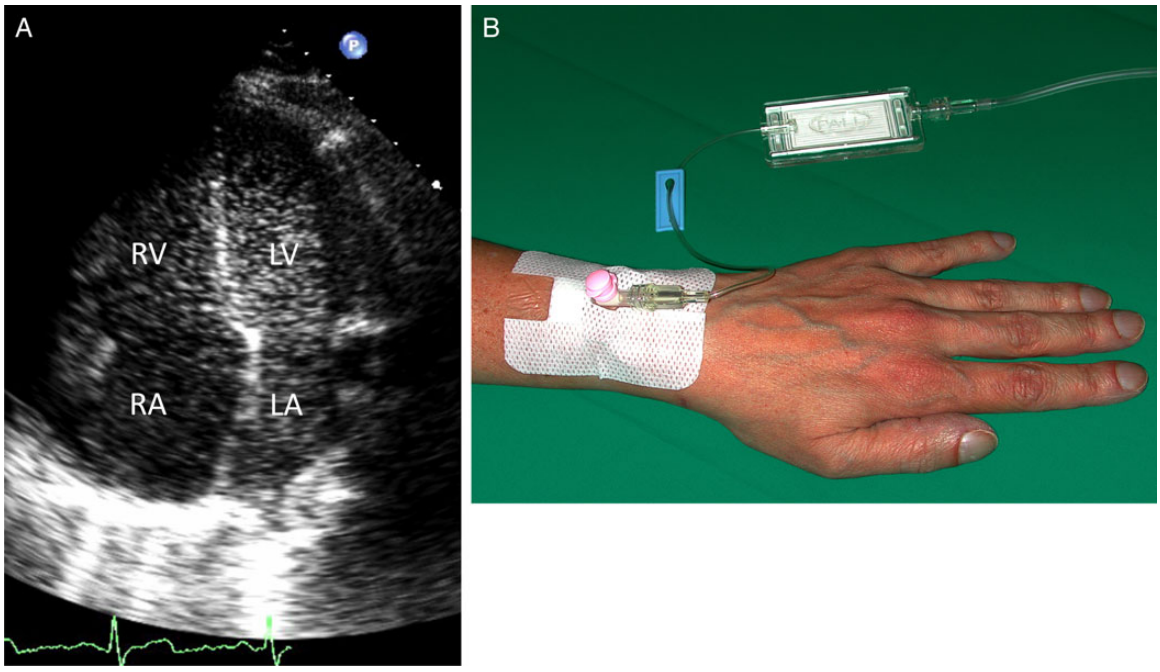

Figure 5 Residual intra-cardiac shunt with risk of paradoxical embolism. (*A*) Echocardiographic four-chamber view after injection of agitated saline contrast in a patient with sinus venosus defect, demonstrating large, spontaneous right-to-left shunting. (*B*) Intravenous line with air filter (bubble trap) to prevent accidental systemic air embolism.

offered prophylactic administration of low-dose aspirin after the 12th week of gestation.

Some commonly used drugs for management of obstetric complications may have adverse effects on the cardiovascular system. Obstetric drugs, their potential cardiovascular side effects, and cautions in their use in women with CHD are listed in *Table 1*.

Multi-disciplinary delivery plan

Haemodynamic changes during delivery and early postpartum in healthy women are depicted in *Figure 2*. The type of CHD and residual haemodynamic lesions may affect the ability of the cardiovascular system to respond to these demands. Effective analgesia and assisted second stage of delivery can effectively decrease the additional haemodynamic load of labour. Assisted delivery may however be associated with a higher risk of postpartum haemorrhage and high-degree lacerations.[65] The average blood loss during vaginal delivery of 500 mL counteracts the impact of auto-transfusion from the contracting uterus at stage 3 of delivery. Blood loss with caesarean section is usually higher (on average 1000 mL). The impact of anaesthesia and anaesthetic drugs on cardiovascular function in women with CHD is beyond the scope of this article but underscores the need for a multi-disciplinary team, including experienced anaesthetists, familiar with special haemodynamic aspects of CHD. With very few exemptions listed in *Box 3*, vaginal delivery, often with early epidural analgesia is the preferred mode of delivery in women with CHD.

A multi-disciplinary delivery plan, based on local experience, logistics, and infrastructure (i.e. possibilities for monitoring on labour ward) should be provided in all women with CHD well before the estimated delivery date. It must be ascertained that this delivery plan is available and accessible at all times and by all team members involved in the care of the woman with CHD. Antibiotic prophylaxis against infective endocarditis is no longer recommended for vaginal delivery. A checklist for important information that must be covered in the delivery plan is outlined in *Box 4*.

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