Pregnancy in adult patients with congenital heart disease

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Abstract

Women with congenital heart disease constitute the majority of cardiac patients encountered in pregnancy, but nevertheless are only infrequently found in the practice of most medical cardiologists and high-risk obstetricians. The patients need guidance regarding contraception and pregnancy, and their various caregivers must share their expertise to promote optimal outcomes. Though maternal mortality is rare except in patients with Eisenmenger syndrome, pulmonary vascular obstructive disease and Marfan syndrome with aortopathy, maternal cardiac complications of pregnancy are more common. As well, women with heart disease are at higher risk for adverse fetal and neonatal outcomes. A review of general and lesion-specific risks and complications of pregnancy and an approach to management are outlined.

Keywords: Pregnancy; Heart diseases, congenital; Prognosis; Management; Contraception

1. Introduction

It is estimated that, in 2000, nearly 60% of persons living with congenital heart disease (CHD) in North America were over the age of 20 (Julien I.E. Hoffman, personal communication); based on current survival data and a relatively constant incidence of new cases, the proportion of CHD patients who are adults will continue to increase for several more decades before it levels off [1]. Women with CHD now comprise the majority of patients with heart disease seen in pregnancy. Nevertheless, they represent a small minority of the pregnant women cared for even in a high-risk obstetrics unit, and they are infrequently encountered by a medical (adult) cardiologist in general cardiology practice. Pediatric cardiologists are, of course, knowledgeable about the underlying cardiac conditions with which such women present, but may be less familiar with pregnancy-related issues. Yet, these women need guidance regarding pregnancy and labor, and contraceptive advice.

Studies of pregnancy outcomes in women with CHD have shown that one can expect a favorable maternal and fetal outcome in most cases. With the exception of patients with Eisenmenger syndrome, pulmonary vascular obstructive disease, and Marfan syndrome with aortopathy, maternal death during pregnancy in women with heart disease is rare [2–6]. However, pregnant women with heart disease do remain at risk for maternal complications including heart failure, arrhythmia and stroke [2–6]. They are also at increased risk for neonatal complications [2,4], a finding we recently confirmed and quantified in a prospective study [7].

2. Cardiovascular physiology and pregnancy

Antepartum, hormonally mediated changes in blood volume, red cell mass, and heart rate result in a 50% increase in cardiac output, increased left ventricular (LV) mass, and decreased peripheral vascular resistance and blood pressure [8]. Changes can be noted by echocardiography by the 14th week of gestation, and peak early in the 3rd trimester [9]. During labor and delivery cardiac output and oxygen consumption increase yet again. Following delivery, relief of caval compression and auto transfusion from the emptied uterus cause a further transient increase in cardiac output. Most of the physiologic changes resolve by the 2nd post-partum week, but complete return to normal may not occur for 6 months [9].
### Table 1
Congenital cardiac lesion in the mother and cardiac risk during pregnancy

<table>
<thead>
<tr>
<th>Risk Level</th>
<th>Lesions</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Low risk</strong></td>
<td>Left-to-right shunts</td>
</tr>
<tr>
<td></td>
<td>Repaired lesions without residual cardiac dysfunction</td>
</tr>
<tr>
<td></td>
<td>Bicuspid aortic valve without stenosis</td>
</tr>
<tr>
<td></td>
<td>Mild-moderate pulmonic stenosis</td>
</tr>
<tr>
<td></td>
<td>Valvular regurgitation with normal ventricular systolic function</td>
</tr>
<tr>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Intermediate risk</strong></td>
<td>Unrepaired or palliated cyanotic CHD</td>
</tr>
<tr>
<td></td>
<td>Uncorrected coarctation of the aorta</td>
</tr>
<tr>
<td></td>
<td>Aortic stenosis (valve area &lt; 1.5 cm² or peak gradient &gt; 30 mmHg)</td>
</tr>
<tr>
<td></td>
<td>Mechanical prosthetic valves</td>
</tr>
<tr>
<td></td>
<td>Severe pulmonic stenosis</td>
</tr>
<tr>
<td></td>
<td>Moderate to severe systemic ventricular dysfunction</td>
</tr>
<tr>
<td></td>
<td>Symptomatic arrhythmia</td>
</tr>
<tr>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>High risk</strong></td>
<td>NYHA functional class III or IV</td>
</tr>
<tr>
<td></td>
<td>Significant pulmonary hypertension with or without right-to-left shunt</td>
</tr>
<tr>
<td></td>
<td>Marfan syndrome with aortic root or major valvular involvement</td>
</tr>
<tr>
<td></td>
<td>Severe aortic stenosis</td>
</tr>
</tbody>
</table>

Modified from Ref. [38].

### 3. Lesion-specific outcomes

#### 3.1. Left-to-right shunts

Pregnancy is generally well tolerated in patients with uncorrected left-to-right shunts, in spite of the volume-loaded right ventricle in atrial septal defect (ASD), or left ventricle in ventricular septal defect (VSD) and patent ductus arteriosus, because the pregnancy-induced decrease in peripheral vascular resistance attenuates the effect of the increase in volume overload [4–6,10]. However, arrhythmias and ventricular dysfunction may occur, especially when the shunt is large. There are anecdotal reports of progression of pulmonary hypertension. The possibility of paradoxical embolization is enhanced when systemic vasodilatation and/or elevation of pulmonary resistance promote transient right-to-left shunting, especially in ASDs.

#### 3.2. Left ventricular outflow tract obstruction

All causes of LV outflow tract obstruction at, below and above the valve have similar hemodynamic consequences in pregnancy. Aortic stenosis secondary to bicuspid aortic valve is the most common of these. In a pregnant woman with severe AS, limited ability to augment cardiac output may result in abnormal elevation of LV systolic and filling pressures that in turn precipitate heart failure or ischemia. In addition the non-compliant, hypertrophiied ventricle is sensitive to falls in preload. The consequent exaggerated drop in cardiac output may lead to hypotension. In a compilation of many small retrospective series, 65 patients were followed through 106 pregnancies with a maternal mortality of 11% and a perinatal mortality of 4% [11]. In 25 of the same 65 pregnancies managed more recently, there was no maternal mortality, although maternal functional deterioration occurred in 20% of pregnancies [11]. Women with symptomatic aortic stenosis should delay pregnancy until after surgical correction [12]. However, the absence of symptoms antepartum is not sufficient assurance that pregnancy will be well tolerated and women with moderate or severe aortic stenosis continue to be at increased risk for pulmonary edema or arrhythmia during pregnancy [6,11] (Table 1). Intrapartum palliation by balloon valvuloplasty may be helpful in selected cases. The possible association of bicuspid aortic valve with ascending aortopathy and coarctation of the aorta should be evaluated.

Patients with well-functioning bioprosthetic aortic valves usually tolerate pregnancy well. It had been claimed in the past that pregnancy might accelerate the rate of degeneration of bioprosthetic or homograft valves, but recent studies have shown that this is not the case [13]. Favorable maternal and fetal outcomes have been reported in women who underwent pulmonary autograft aortic valve-replacement (Ross procedure) [14]. Pregnancy in women with mechanical valve prostheses carries increased risk (3–14%) of valve thrombosis, especially if subcutaneous unfractionated heparin rather than warfarin is used as the anticoagulant. (See later discussion of anticoagulation in Section 4.2.)

#### 3.3. Coarctation of the aorta

In early series, uncorrected coarctation was associated with 3% maternal mortality, and the risk was greater in the presence of associated cardiac defects, aortopathy, or long-standing hypertension. Recent studies of corrected and uncorrected patients have been more encouraging, with only one maternal death reported in 182 pregnancies [15]. The death occurred in a woman who had previously undergone coarctation repair, emphasizing that while prior repair reduces maternal risk, it does
not eliminate it [5,6,15]. Pregnant women with repaired coarctation are at increased risk for pregnancy-induced hyperten-
sion [5,6,15], likely as a result of residual abnormalities in aortic compliance. Satisfactory control of upper body hypertension during pregnancy may lead to excessive hypotension below the coarctation site, compromising the fetus. In unrepaird coarctation, the incidence of intrauterine growth restriction and premature labor are increased.

3.4. Pulmonary stenosis

Mild pulmonic stenosis or pulmonic stenosis that has been alleviated by valvuloplasty or surgery is well tolerated during pregnancy and fetal outcome is favorable [5,6]. In severe pulmonic stenosis the increased hemodynamic load of pregnancy may precipitate right heart failure or atrial arrhythmias even in a previously asymptomatic woman, so correction prior to pregnancy should be considered. Balloon valvuloplasty can be accomplished during pregnancy if symptoms develop in a previously untreated patient.

3.5. Cyanotic heart disease: unrepaired and repaired

Patients with cyanotic CHD such as previously unoperated or palliated tetralogy of Fallot, single ventricle, etc., are subject to increased right-to-left shunting, hence increased maternal hypoxemia and cyanosis, because of the pregnancy-associated fall in systemic vascular resist-
tance. Forty-four women with a variety of cyanotic congenital heart defects who were followed through 96 pregnancies were shown to have a high rate of maternal cardiac events (32% including 1 death), prematurity (37%) and a low live birth rate (43%) [16]. When the maternal oxygen saturation was \( \leq 85\% \), the live birth rate was only 12%.

After correction of tetralogy of Fallot, pregnancy risk is low [4–6] but may be increased depending on residua and sequelae such as residual shunt, right ventricular outflow tract obstruction, arrhythmias, pulmonary regur-
gitation, right ventricular systolic dysfunction, pulmo-
nary hypertension, or LV dysfunction.

Women who have had an atrial repair (i.e. Mustard or Senning procedure) for complete transposition of the great arteries are subject to late adult complications such as sinus node dysfunction, atrial arrhythmias, and dys-
function of the systemic ventricle. In 43 pregnancies in 31 women described in recent reports, there was 1 late maternal death [17,18]. There was a 14% incidence of maternal heart failure, arrhythmias, or cardiac deterior-
ation. No studies of pregnancy outcome in women who received the current repair of choice for complete trans-
position, the arterial switch procedure, have yet been reported. However, in the absence of ventricular dys-
function, coronary obstruction, or severe valve dysfunc-
tion, a good outcome is expected.

Although the Fontan operation eliminates cyanosis and volume overload of the functioning systemic ven-
tricle, patients have a limited ability to increased cardiac output. Reported data on pregnancy outcomes include only patients who were doing well prior to pregnancy. In 21 such women, only 15 of 33 pregnancies (45%) continued to completion. Two women had cardiac comp-
lications and the incidence of first trimester miscarriage was high (39%), but there was no maternal mortality in this selected series [19].

3.6. Congenitally corrected transposition of the great arteries

Many adult patients will have had surgical interven-
tions, primarily VSD closure and relief of pulmonic stenosis, sometimes requiring a valved conduit from the LV to the pulmonary artery. Potential problems in pregnancy in women with congenitally corrected trans-
position include dysfunction of the systemic right ven-
tricle and/or increased systemic AV valve regurgitation with heart failure, atrial arrhythmias, AV block, and sequelae of prior surgical interventions for associated anomalies. In two recent reports on 41 patients, there were 105 pregnancies with 73% live births. No maternal mortality occurred in these studies, but 7 patients developed heart failure, endocarditis, stroke, or myocardial infarction [20,21].

3.7. Marfan syndrome

The risk of life-threatening aortic complications of Marfan syndrome is increased in pregnancy due to hemodynamic stress and perhaps hormonal effects. Patients entering pregnancy with a normal aortic root size and no prior aortic surgery usually do well, though dissection can occur without pre-existing aortic abnor-
malities. A prospective study of 45 pregnancies in 21 patients reported no increase in obstetrical complications or significant change in aortic root size in patients who began the pregnancy with normal aortic roots. However, in the 8 patients with a dilated aortic root (>40 mm) or prior aortic root surgery, 3 of 9 pregnancies were complicated by either aortic dissection (2) or rapid aortic dilatation (1) [22]. Thus, patients with aortic root involvement should receive preconception counselling emphasizing their risk, and in early pregnancy should be offered termination. The likelihood of aortic dilatation increases with increasing maternal age, so advice to complete families at a younger age is appropriate. Serial echocardiography should be used to identify progressive aortic root dilatation during pregnancy and for 6 months post-partum. Prophylactic beta-blockers should be administered [23]. The aortopathy in Marfan syndrome
is a generalized process. Patients who already demonstrate root dilatation likely have more severe aortic pathology than those whose ascending aortic dimension is still normal, hence may not eliminate their increased risk of dissection of the residual native aorta by undergoing prophylactic root replacement prior to pregnancy.

3.8. Eisenmenger syndrome and pulmonary vascular obstructive disease

Maternal mortality in Eisenmenger syndrome is approximately 30% in each pregnancy [24]. The preponderance of complications occurs at term and during the first post-partum week. Preconception counselling should stress the extreme pregnancy-associated risks. Termination of pregnancy should always be offered to such patients, as should sterilization. Further, fetal outcomes are poor, due in part to increasing right-to-left shunting as a result of pregnancy-induced fall in peripheral vascular resistance, and consequent worsening of maternal cyanosis. Spontaneous abortion, intrauterine growth restriction (30% of pregnancies), and pre-term labor are common, with a high perinatal mortality rate (28%), due mainly to prematurity.

In a recent review of the outcome of 125 pregnancies in patients with Eisenmenger syndrome, primary pulmonary hypertension and secondary pulmonary hypertension [25], maternal mortality was 36, 30 and 56% respectively. The overall neonatal mortality was 13%.

4. Management

4.1. Risk stratification and counselling

Issues regarding contraception and pregnancy should be discussed as soon as patients reach an age when they may become sexually active; thus pediatric cardiologists often have the first opportunity to provide preconception counselling. The data required for pregnancy risk stratification can be acquired readily from a thorough cardiovascular history and examination, review of prior records, 12-lead electrocardiogram, transthoracic echocardiogram, and arterial oxygen saturation when indicated.

The cardiologist plays his or her most important role in defining the underlying cardiac lesion and the nature of residua and sequelae, especially ventricular function, pulmonary pressure, severity of obstructive lesions, persistence of shunts, and presence of hypoxemia. Often, such information is impossible for other caregivers, such as anesthetists and obstetricians, to access and evaluate, absent a clear exposition by the cardiologist.

Classically, patients have been stratified by the nature of their lesion into low-, intermediate-, or high-risk groups (Table 1). Maternal status defined by New York Heart Association (NYHA) functional class has been used traditionally as a predictor of outcome. In an older study of 482 pregnancies in women with CHD, cardiovascular morbidity was less (8% vs. 30%) and live birth rate higher (80% vs. 68%) in mothers with NYHA functional class I compared to the others [2]. In two contemporary studies examining the outcomes of 851 pregnancies, poor functional status (NYHA > II) or cyanosis, LV systolic dysfunction, left heart obstruction, and history of cardiac events prior to pregnancy (arrhythmia, stroke, or pulmonary edema) were independent predictors of maternal cardiac complications [5,6]. Poor maternal functional class or cyanosis was predictive also of adverse neonatal events.

A ‘risk index’ has been developed, incorporating these maternal risk factors, which are definable prior to or early in pregnancy. The risk of a cardiac event (cardiac death, stroke, pulmonary edema, or arrhythmia) during pregnancy increased with the number of predictors present during the antepartum evaluation. In a woman with heart disease and a risk index of 0, the likelihood of a cardiac event during pregnancy is approximately 5%, whereas with a risk index of 1 it rises to 27%, and with a risk index > 1, the likelihood is 75% [6]. Although the risk appears alarmingly high, most of the morbid events are nonfatal. There was no cardiac mortality in the group with risk score = 0, and the cardiac mortality rate in pregnancies with risk score ≥ 1 was 1%. It is important to use this index together with lesion-specific risk estimates where available, since certain populations with known lesion-specific risks are not defined by the global risk index. This is at least partially because patients with previously established high risk (e.g. Marfan with dilated root, Eisenmenger), were under-represented in the contemporary population of pregnant women with heart disease from which the global risk index was derived (Table 2).

Women with heart disease have an increased risk of neonatal complications [2–7]. In a recently completed prospective study, we showed that the risk of neonatal complications (premature birth, small-for-gestational-age birth weight, respiratory distress syndrome, intraventricular hemorrhage, fetal or neonatal death) is amplified if there are concomitant maternal non-cardiac (obstetrical and other) risk factors for neonatal complications [7] (Table 2).

Women with a high risk score (≥ 1) for cardiac complications [6], those with lesion-specific risks, and those at risk for neonatal complications [7] should benefit from enhanced multidisciplinary surveillance in a high-risk obstetrics unit, whereas those with no such risk factors may do well with normal obstetrical and cardiac care in the community.

Both maternal and fetal outcomes are improved by surgery to correct cyanosis, which should be undertaken prior to conception when possible [4]. Similarly, patients with symptomatic obstructive lesions should undergo
Table 2

Risk factors for maternal cardiac and neonatal adverse events in pregnancy in women with heart disease

<table>
<thead>
<tr>
<th>Nature of risk</th>
<th>Risk factor</th>
</tr>
</thead>
</table>
| Maternal cardiac adverse event (pulmonary edema, arrhythmia, stroke, death) | Poor functional class (NYHA Class>II) or cyanosis$^a$
Systemic ventricular systolic dysfunction (EF <0.40)$^a$
Left heart obstruction (mitral valve area <2.0 cm$^2$, aortic valve area <1.5 cm$^2$, or peak LVOT gradient >30 mmHg)$^a$
Cardiac event (arrhythmia, stroke, TIA, pulmonary edema) prior to pregnancy$^a$
Known lesion-specific risk |
| Neonatal adverse event (premature birth, small-for-gestational-age birth weight, respiratory distress syndrome, intraventricular hemorrhage, fetal or neonatal death) | Presence of maternal heart disease (increased risk amplified by the additional presence of risk factors listed below)
Poor maternal functional class (NYHA Class>II) or cyanosis
Maternal left heart obstruction
Maternal age <20 or >35 years old
Obstetric risk factors for adverse neonatal events$^b$
Multiple gestation
Smoking during pregnancy
Anticoagulant therapy |

$^a$ The risk factors marked (a) may be used to constitute a risk index: the risk of a maternal cardiac adverse event with 0 risk factors present is <5%, with 1 risk factor present is 25%, and with more than one risk factor present is 75% (from Ref. [6]).

$^b$ History of premature delivery or rupture of membranes, incompetent cervix, or caesarean section; or intrauterine growth retardation, antepartum bleeding >12 weeks gestation, febrile illness, or uterine/placental abnormalities during present pregnancy.

intervention prior to pregnancy [12]. During pregnancy, the result of cardiovascular surgery is less favorable, with maternal and fetal mortality of 6 and 30%, respectively [26]. The lack of ideal choices once severe valve disease is present argues for completing families earlier, before age-dependent progression necessitates valve-replacement surgery and raises the difficult issues of anticoagulation required by mechanical valves, or predictable need for earlier re-operation if a tissue valve is used.

Additional associated risk factors that may complicate pregnancy include a history of prosthetic valves and conduits, anticoagulant therapy, and the use of teratogenic drugs such as warfarin or angiotensin converting enzyme inhibitors.

The issue of limited maternal long-term prognosis is sometimes first addressed in the context of pregnancy counselling. Information regarding life expectancy must be shared to allow the prospective mother and her family to make an informed decision. Similarly, when the maternal condition imparts a high likelihood of fetal complications, (e.g. a mother with cyanosis or on anticoagulants), this must be explained.

The risk of recurrence of CHD in offspring should be discussed in the context of a 0.4–0.6% risk in the general population [27]. The risk with a first-degree relative affected increases approximately 10-fold overall, but is higher in left heart obstructive lesions. Autosomal dominant conditions such as Marfan syndrome and the 22q11 deletion syndromes confer a 50% risk of recurrence in an offspring. Formal genetic counseling is helpful, to ensure that patients are fully informed of the mode of inheritance, recurrence risk, prenatal diagnosis options available and preventive strategies to decrease the incidence of congenital defects, for instance preconception use of multivitamins containing folic acid [28].

4.2. Antepartum management

Pregnant women with heart disease may be at particular risk for one or more of congestive heart failure; arrhythmias; or thrombosis, emboli, and adverse effects of anticoagulants.

Activity limitation is helpful in symptomatic women with ventricular or valvular dysfunction, and the most severely limited may need hospital admission if symptoms progress in spite of therapy. Gestational hypertension, hyperthyroidism, infection and anemia should be identified early and treated vigorously. For patients with functionally significant mitral stenosis, beta-adrenergic blockers rather than digoxin should be used to control heart rate. We also offer empiric therapy with beta-adrenergic blockers to patients with coarctation and to Marfan patients.

4.2.1. Arrhythmias

Pregnancy may exacerbate the frequency and hemodynamic severity of arrhythmias in those who have experienced them prior to pregnancy; arrhythmias may also arise de novo during pregnancy. Pharmacological treatment should be reserved for patients with severe symptoms or when sustained episodes are poorly tolerated in the presence of ventricular hypertrophy, ventricular dysfunction, or valvular obstruction. Sustained tachyarrhythmias such as atrial flutter or atrial fibrillation should be treated promptly, avoiding teratogenic
drugs. Digoxin and beta-adrenergic blockers are drugs of choice for this purpose in view of their known safety profiles [29]. Quinidine, adenosine, sotalol and lidocaine are also generally accepted as ‘safe’. There are case reports describing successful use of amiodarone though it is classified as contra-indicated in pregnancy in standard texts. It is not teratogenic, but may impair neonatal thyroid function [30,31]. External electrical cardioversion is safe in pregnancy. There is a report of 44 pregnancies in patients with implantable cardioverter-defibrillators, with favorable maternal and fetal outcomes [32].

4.2.2. Anticoagulation

There is no perfect anticoagulation strategy during pregnancy, and controversy about the optimal strategy will not be resolved without clinical trials. Oral anticoagulation with warfarin is effective and logistically easier. Warfarin embryopathy may be produced during organogenesis, though there is some evidence that a daily warfarin dose of \( \leq 5 \text{ mg} \) may not be teratogenic [33]. Fetal intracranial bleeding can occur throughout pregnancy when the mother takes warfarin, and is a particular risk during vaginal delivery unless warfarin has been stopped at least two weeks prior to labor. Adjusted dose subcutaneous heparin has no teratogenic effects, as the drug does not cross the placenta, but heparin may cause maternal thrombocytopenia and osteoporosis. Claims of inadequate effectiveness of heparin in patients with mechanical heart valves have been countered by arguments that inadequate doses were used. In a systematic overview of prior studies examining the relationship of anticoagulation regimen and pregnancy outcomes in women with prosthetic heart valves have been countered by arguments that inadequate doses were used. In a systematic overview of prior studies examining the relationship of anticoagulation regimen and pregnancy outcomes in women with prosthetic heart valves have been countered by arguments that inadequate doses were used.

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4.2.3. Eisenmenger syndrome

If a woman with Eisenmenger syndrome does not accept counselling to terminate, or presents late in pregnancy, meticulous antepartum management is necessary including early hospitalization, supplemental oxygen and possibly empiric anticoagulation. The efficacy of nitric oxide therapy in these patients has yet to be demonstrated.

4.3. Multidisciplinary approach and high-risk pregnancy units

Women with heart disease who are at intermediate or high risk for complications (Tables 1 and 2) should be managed in a high-risk pregnancy unit by a multidisciplinary team from obstetrics, cardiology, anesthesia and pediatrics. When dealing with a complex problem the team should meet early in the pregnancy. At this time the nature of the cardiac lesion, anticipated effects of pregnancy, and potential problems should be explored. Since it is often not possible for every member of the team to be at the patient’s bedside at a moment of crisis, it is helpful to develop and distribute widely a written management plan for foreseeable contingencies. Women with heart disease at low risk can be managed in a community hospital setting [6,7]. However, if there is doubt about the mother’s status or the risk, consultation at a regional referral centre should be arranged.

4.4. Labor and delivery

The only definite cardiac indications for caesarean section are aortic dissection, Marfan syndrome with dilated aortic root, and failure to switch from warfarin to heparin at least 2 weeks prior to labor. Otherwise, vaginal delivery is preferred. Pre-term induction is rarely indicated, but once fetal lung maturity is assured a planned induction and delivery in high-risk situations will ensure availability of appropriate staff and equipment. Although there is no consensus on the use of invasive hemodynamic monitoring during labor and delivery, we commonly utilize intra-arterial monitoring and often use central venous pressure monitoring as well in cases where there are concerns about the interpretation and deleterious effects of a sudden drop in systemic blood pressure (e.g. patients with severe aortic stenosis, pulmonary hypertension, severe systemic ventricular systolic dysfunction, or preload dependent physiology such as Fontan). The clinical utility of an indwelling pulmonary artery catheter has not been studied in pregnancy. A PA catheter is utilized, rarely, in situations where the information sought is not available otherwise and warrants the risk of the procedure, considering also that the risk of its insertion may be increased because of complex anatomy such as atrial baffles, or in the setting of pulmonary hypertension because of possible pulmonary infarction or rupture.

Heparin anticoagulation is discontinued at least 12 h prior to induction, or reversed with protamine if spon-
taneous labor develops, and can usually be resumed 6–12 h post-partum. Many centres with extensive experience in caring for pregnant women with heart disease utilize endocarditis prophylaxis routinely but there is no evidence to support this common practice, which is not recommended for cesarean section delivery or for uncomplicated vaginal delivery in absence of infection according to current American Heart Association guidelines [37]. If unanticipated bacteremia is suspected during vaginal delivery, intravenous antibiotics can be administered at the time [37].

Epidural anaesthesia with adequate volume preloading is the technique of choice. Epidural fentanyl is particularly advantageous in cyanotic patients with shunt lesions or those with significant aortic stenosis, as it does not lower peripheral vascular resistance. In the presence of a shunt, air and particulate filters should be placed in all intravenous lines.

Labor is conducted in the left lateral decubitus position to attenuate hemodynamic fluctuations associated with contractions in the supine position. Forceps or vacuum extraction will shorten the latter part of the second stage of labor and reduce need for maternal expulsive efforts. As hemodynamics do not approach baseline for many days after delivery, patients at intermediate or high risk may require monitoring for a minimum of 72 h post-partum. Patients with Eisenmenger syndrome require longer close post-partum observation, since mortality risk persists for 7 days or more.

5. Contraception

Contraceptive planning is an important part of the counselling process in general, and should be reviewed again early post-partum. Choice of method should take into account the efficacy and risks of each method, patient preference, and the consequences of contraceptive failure (Table 3).

Table 3
Contraception in patients with heart disease

<table>
<thead>
<tr>
<th>Type of contraceptive</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oral contraception (combined estrogen and progesterone)</td>
<td>Contra-indicated where there is a risk of thrombotic events and/or hypertension. Should be used with caution if cardiac reserve is marginal. May be used in patients already on anticoagulants for other reasons. Pregnancy rate &lt;1% per year.</td>
</tr>
<tr>
<td>Progestin only</td>
<td>Short-term method of choice in many patients at risk of thrombotic events, but have a higher failure rate than combined estrogen–progesterone compounds, and more side effects. In combination with breast-feeding the progestin only mini-pill provides excellent efficacy.</td>
</tr>
<tr>
<td>Intrauterine device</td>
<td>Contra-indicated where there is a risk of endocarditis. Pregnancy rate 3% per year.</td>
</tr>
<tr>
<td>Barrier methods</td>
<td>In many instances the method of choice for short-term family planning. The most effective of these, condom plus spermicidal foam yields a pregnancy rate of 3% per year in compliant users.</td>
</tr>
<tr>
<td>Tubal ligation</td>
<td>In the presence of significant cardiac disease, method of choice once family is complete. Optimal approach if further pregnancies are contra-indicated because of cardiac status.</td>
</tr>
</tbody>
</table>

Modified from Ref. [39].

References


