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Editorial

Pregnancy and Cardiac Disease

Embarazo y enfermedad cardiaca

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Cardiovascular disease (CVD) complicating pregnancy is an increasingly frequent phenomenon with regional differences in numbers and causes and substantial changes over the last few decades. Between 0.2% to 4% of pregnancies are affected in countries with a developed health care system.[1] Cardiac disease in the mother is the most important cause of maternal mortality and there have been no signs of decline in its incidence over the past few decades. On the contrary, there has been an increase, which is well documented for the United Kingdom and the United States. Aortic dissection, peripartum cardiomyopathy, and ischemic heart disease are rare but important causes of maternal mortality. The prevalence of ischemic heart disease has increased due to life style changes with more women with obesity, hypertension, and diabetes. In addition, the tendency to older age at first pregnancy, including the progressive rise in in vitro fertilization and multiple pregnancies, has increased the prevalence of CVD.

One of the other major causes of CVD complicating pregnancy is the dramatic increase in patients with congenital heart disease (CHD) surviving to adulthood. In the 1950s only around 10% survived the first decades of life whereas today up to 85% to 90% reach adulthood.[2] These women not only survive but, due to the tremendous advances in the diagnosis and treatment of even complex CHD, their quality of life has improved, enabling them to lead a normal life, including sexual activity and the desire for children. A studyconducted in the United States[3] showed that, from 2000 to 2010, there was a linear increase in the prevalence of CHD from 6.4 to 9.0 per 10 000 of delivery hospitalizations and, furthermore, a mortality rate of 17.8 compared with 0.7/10 000 delivery hospitalizations in mothers without CHD. Like other investigations, that study showed the predominance of patients with an underlying left-to-right shunt lesion (atrial septal defect in 22.6%, ventricular septal defect in 14.5%) followed by valvular lesions (left-sided in 13.8%, right-sided in 7.5%). More complex lesions such as tetralogy of Fallot (2.6%) or transposition of the great arteries (1.5%) were seen only rarely.

While many women with known CVD, whether treated or not, may be in a stable condition, pregnancy, with its significant hemodynamic changes, puts them at risk for deterioration. Stroke volume, heart rate, and cardiac output increase, whereas peripheral vascular resistance and hemoglobin decrease. All changes occur slowly, with a peak effect in the late second and early third trimester, normalizing only weeks after delivery. In addition to the hemodynamic effects, pregnancy has a thrombogenic effect and may affect connective tissue disease leading to a higher risk of aortic dissection during pregnancy. Therefore, pregnancy planning and contraceptive advice are essential for women with cardiac disease.[4] Prior to pregnancy, all women with known CVD should be seen and counselled by well-trained specialists in a center with expertise. An experienced multidisciplinary team should be available to provide care before, during, and after pregnancy.[5]

Women with preexisting CVD are known to be at higher risk of developing cardiac complications in pregnancy, which are strongly influenced by the kind of heart defect and residual lesions. Efforts have been made in the field of risk assessment and stratification. The modified World Health Organization (mWHO) classification seems to be the most accurate measure in predicting these risks.[6] It divides patients based on their underlying diagnosis in 4 groups from very low risk (mWHO I) to those in whom pregnancy poses a life-threatening risk and is therefore contraindicated (mWHO IV).[7] The low-risk group (mWHO I) is composed of all patients with minor lesions not requiring treatment and those that have been treated without hemodynamically significant residuals. Pregnancy is contraindicated (mWHO IV) in women with pulmonary hypertension, severe cyanosis, diminished left ventricular function, previous peripartum cardiomyopathy with incomplete recovery, symptomatic left ventricular outflow tract obstruction, and Marfan syndrome with aortic root dilation.

For a better understanding and management of women with CVD, guidelines have been developed covering most of the relevant issues.[1] The guidelines are based on the available literature, which consists mainly of reports from single centers in different parts of the world and using distinct methods. They often focus on 1 lesion only and describe small groups of patients. Therefore, it used to be difficult to obtain reliable and relevant information.

To gather up-to-date information about treatment during pregnancy and pregnancy outcomes, a large prospective observational registry has been initiated by the European Society of Cardiology and Association for European Paediatric and Congenital Cardiology: the Registry of Pregnancy and Cardiac Disease (ROPAC), which is web-based and open for inclusion to all

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cardiologists and obstetricians managing patients with heart disease. Currently, 132 centers from 42 countries on all continents contribute data to this prospective registry, which has included over 4000 pregnancies to date. The goals of ROPAC are to assess the risks to both mother and child, to outline the variation between different regions and parts of the world, to test and validate existing risk models, to provide better knowledge for guidelines, and finally to provide optimal advice to women and improve their management and outcomes. Inclusion criteria are pregnant women with structural heart disease, whether congenital or acquired, cardiomyopathies, aortic pathology, or pulmonary hypertension. Exclusion criteria consist of the absence of structural heart disease such as dysrhythmias. All consecutive patients with structural heart disease diagnosed before pregnancy are enrolled, and data are recorded on the current pregnancy and a 6 month follow-up period. Several articles and reports from this registry have been published.[7,8,10–15] Most patients in ROPAC have CHD (52%), followed by valvular disease (32%), cardiomyopathy (7%), aortic disease (3%), ischemic heart disease (1.5%), and pulmonary hypertension (0.5%). In developed countries, 74% of the women had CHD, while in developing countries 72% had acquired valvular heart disease.

The most frequently encountered complications are heart failure and arrhythmias. Heart failure occurred in 13% of patients in ROPAC, mainly at the end of the second trimester or shortly after delivery. The patients found to be especially at risk were those with an episode of heart failure before pregnancy and those with a diagnosis of cardiomyopathy.[9]

Arrhythmias are also an issue in the cardiac management of patients with heart disease.[10] The most common arrhythmias are supraventricular in origin. Chronic hemodynamic stress may cause atrial dilatation and subsequent atrial arrhythmias. Scar tissue and suture lines created by corrective surgery may function as pathways for macroreentry within atrial tissue (intra-atrial reentrant tachycardia). Supraventricular ectopy and supraventricular tachycardias are often seen in healthy pregnant women but atrial fibrillation or flutter are very rare. In ROPAC, atrial fibrillation or flutter during pregnancy in women with heart disease was associated with a marked increase in maternal mortality and low birth weight. Atrial fibrillation/atrial flutter occurred in 1.3% of the patients with structural heart disease, reaching a peak between weeks 23 and 26 of pregnancy.[11] Ventricular tachyarrhythmias (VTA) may either occur by macroreentry pathways after surgery involving the ventricles or by ventricular dysfunction after long-standing hemodynamic stress. These tachyarrhythmias occurred in 1.4% of pregnant women with CVD in ROPAC, mainly in the third trimester, and were associated with heart failure during pregnancy. The New York Heart Association class before pregnancy was predictive. Ventricular tachyarrhythmias during pregnancy had a clear impact on fetal outcome: neonatal death, preterm birth (< 37 weeks), low birthweight (< 2500 g) and Apgar score < 7 were more common in women with VTA.[12] The use of medication is more complex during pregnancy because some drugs, such as vitamin K antagonists or angiotensin-converting enzyme inhibitors, may have harmful effects on the fetus, while others may influence birthweight, such as beta-blockers.[13,14]

Patients with a mechanical prosthesis are a specific high risk group. The anticoagulation regimen is difficult during pregnancy, especially because pregnancy is a thrombogenic situation. Vitamin K antagonists are most effective in preventing valve thrombosis and are probably the best option for the mother, but at the cost of fetal abnormalities and a higher miscarriage rate. Heparin is safe for the fetus but is associated with valve thrombosis. In ROPAC, 212 women with a mechanical prosthesis were studied and only 58% delivered a live newborn without serious complications.[15] Therefore, it may be wise to refer these patients to a specialized center.

Finally, in patients with heart disease, the mode of delivery should be discussed in a timely fashion by a multidisciplinary team. Vaginal delivery is the preferred mode of delivery in most patients, with epidural anesthesia and an assisted second stage, when needed.[16] A cesarean section is preferred only in some high risk groups, such as patients with a dilated aorta or severe heart failure.[1]

Because larger numbers are still needed to obtain reliable information on specific subgroups, ROPAC will continue to recruit new centers and expand the numbers of patients in the coming years to enroll at least 10 000 women with cardiac disease. Centers wishing to be recognized as specialized centers and to be able to participate in trials and registries must meet the recommendations specified in the position paper of the Working Group on Grown-up CHD of the European Society of Cardiology. It is essential to adopt a multidisciplinary approach, with the co-located presence of pediatric and adult cardiology and surgery forming a congenital cardiology team, with close collaboration with the department of obstetrics and gynecology.[5] In addition to large prospective registry data, specific issues, such as the type of anticoagulation during pregnancy, warrant the setting up of randomized trials, although this may be difficult to achieve during pregnancy.

CONFLICTS OF INTEREST

None declared.

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