Editorial

Management of pregnancy in women with congenital heart disease

The number of women with congenital heart disease reaching childbearing age is increasing, and this group now represents the majority of women with heart disease during pregnancy. This increase raises certain issues which have already been addressed in editorials in this journal, relating to risk factors and the difficulties arising from pregnant women with severe congenital heart disease.

Simple defects
Atrial and ventricular septal defects, and arterial duct, when operated on early in infancy, do not require any special treatment. Asymptomatic left-to-right shunts and moderate pulmonary stenosis are also usually well tolerated. These women should be able to give birth in non-specialist centres. The only concern is prevention of endocarditis in case of residual shunts or valvar regurgitation. Atrial septal defect is the most common congenital heart disease in adults, and in the absence of pulmonary hypertension induces barely any complications.

Contraindications
Eisenmenger reaction, major or significant aortic dilatation in Marfan syndrome, severe aortic stenosis, deep cyanosis, and heart failure put the patient at high risk of maternal death, even where there is close follow up. Eisenmenger reaction carries up to 50% mortality. Recent inquiries in the UK report maternal death in the range of 2-11/100,000 from 1994-96. The main cardiac causes of death are pulmonary hypertension or dissecting aneurysm.

Recommendations for Eisenmenger patients include avoidance of vasodilatation, maintenance of preload, and use of prophylactic heparin. Marfan disease with aortic dilatation should contraindicate pregnancy when the ascending aorta is more than 40 mm as there is an increasing risk of aortic dissection. Even if the aorta is smaller, pregnancy should be followed up closely with monthly echocardiograms.

Palliated or balanced heart defect
Difficult issues can also arise in women who have undergone palliative surgical procedures. These patients lives are not usually at risk but they may suffer morbidity and the fetus may be harmed.

Understanding the physiological changes induced by pregnancy can help to anticipate or prevent complications: an increase in blood volume (40%), a fall in peripheral resistance, an increase in cardiac output (40%), and an altered heart rate are the most important of these changes. Potential for thrombosis is high during pregnancy and prolonged several weeks after delivery. Labour and delivery cause the blood volume and systemic resistance to vary greatly.

Pregnant women with congenital heart disease can be divided into five groups:
(1) The systemic ventricle is the right ventricle, as in transposition of the great arteries having undergone an atrial switch, or in double discordance. Adding to potential arrhythmias induced by surgery, the risk is deterioration of the right ventricular function with increasing atrioventricular regurgitation, sometimes irreversible after pregnancy. Several studies have reported good outcomes of pregnancy in selected women with atrial switch and with double discordance.

(2) Overloaded right ventricle caused by pulmonary regurgitation and/or obstruction. This is often the case in the following situations: operated tetralogy of Fallot, right ventricle to pulmonary conduits used to repair tricuspid or pulmonary atresia with ventricular septal defect (VSD), or repaired malposition of the great arteries with VSD. This situation is often well tolerated, even in cases of severe pulmonary regurgitation, and delivery might be normal. The fear of ventricular or atrial arrhythmia can sometimes lead to pre-pregnancy pulmonary valve replacement, if the functional status before pregnancy is greatly impaired.

(3) Cyanotic patients with Ebstein anomaly, palliated pulmonary atresia with VSD, univentricular heart physiology, excluding Eisenmenger reaction. Vasodilatation induced by pregnancy usually increases cyanosis and can cause the mother’s condition to deteriorate. In addition, hypoxaemia influences fetal growth and is linked to a high incidence of miscarriage when oxygen saturation is less than 85% or when haemoglobin is more than 18 g/dl (11.2 mmol/l). A moderate cyanosis increases the incidence of low birth weight and premature birth, as well as the risk of maternal thromboembolic events.

(4) Moderate alteration of left ventricular function. Careful evaluation cannot always formally predict risk. Regular review should be undertaken during pregnancy, and early delivery should be considered if there is progressive deterioration in left ventricular function.

(5) Residual aortic coarctation or moderate left outflow tract obstruction. In case of residual coarctation a high level of upper limb systemic pressure should not induce inappropriate medical treatment for hypertension because of possible impairment of blood supply to the placenta. Patients with moderate left sided outflow tract obstruction benefit from rest in hospitals.

For all these patients obstetricians, anaesthetists, and cardiologists should closely cooperate, ideally on the same site. Maternal cardiac risks are mainly atrial arrhythmias (groups 1, 2, and 3), cardiac failure (groups 1, 3, and 4), increasing cyanosis (group 3), and thromboembolic events (groups 3 and 4).

Management
PRECONCEPTION OUTPATIENT CLINIC
It is essential to:
- inform the patient about the risks as far as one can anticipate them;
- assess the functional ability with echocardiography and exercise testing before pregnancy;
- carefully consider the potential risk to the fetus of maternal medication with agents such as angiotensin.
Recent data suggest that
is an exception because of the risk of fetal hypothyroidism.

- encourage pregnancy early in life;
- inform the patient about the recurrent risks of congeni-
tal heart defects (as high as 6% in tetralogy of Fallot11);
- inform the patient about prenatal diagnosis, not only of
heart defects but also about associated problems such as
 genetic issues;
- consider giving multivitamins to the patients three
months before pregnancy and during the first three
months after—these have been proven to lower the inci-
dence of fetal heart defect in the general population12;
- detect risk factors for main obstetric complications (pre-
ceclampsia, early preterm delivery, fetal death in utero).

DURING PREGNANCY
Clinical assessment associated with regular comparative
echocardiograms is wise for all patients, but follow up
requires an individual approach for each patient, especially
in the presence of impaired right ventricular function. Fetal
heart scanning is traditionally offered at 18 weeks. Fetal
growth, especially in cyanotic women, has to be carefully
monitored.

Questions from obstetricians should be addressed. Can
the patient have:
- tocolysis by β stimulation or calcium channel inhibitors? Yes, but this may result in hypotension
- steroids (at high doses) for pulmonary maturation in
case of premature labour to avoid fetal pulmonary
distress? Yes, but they can induce volume expansion
- elective induced labour with oxytocic or prostaglandin?
Yes, but there is a risk of bradycardia, hypertension,
myocardial ischaemia, and vasospasm
- spinal anaesthesia? Yes, but since volume expanding
solutions will be given, the risk of cardiac failure should
be compared to the benefit of analgesia which will mini-
mise haemodynamic changes during labour
- arterial pressure monitoring during labour (in case of
sacrificed subclavian arteries)? Yes, but via an intravas-
cular line, if necessary
- antibiotic prophylaxis? For normal vaginal delivery it is
not recommended by the American Heart Association.13
Although well documented cases of endocarditis are
uncommon, they can be very severe and be prophy-
laxis (ampicillin) at the start of the labour and up to 48
hours
- planned breastfeeding? Yes, if the mother is not given
drugs that are dangerous for the newborn

TREATING COMPLICATIONS
Most antiarrhythmic drugs are well tolerated and can be given
at relatively low risk to treat atrial arrhythmias.14 Amiodarone
is an exception because of the risk of fetal hypothyroidism.
Recent data suggest that β blockers may cause intrauterine
growth retardation if they are administered during the first
trimester. In complex heart defects with “fragile ventricles”,
the use of potentially proarrhythmic drugs should be
avoided. Direct current cardioversion to terminate maternal
arrhythmias is well tolerated and effective.

There is no consensus about use of anticoagulants in
cyanotic patients but low molecular weight heparin can
only be a good alternative to oral anticoagulation. In
patients with mechanical valves, the classical scheme is not
ideal. Subcutaneous heparin during the first trimester,
while avoiding teratogenicity associated with an oral
anticoagulant, carries the risk of thrombosis. Switching
from oral anticoagulant to subcutaneous heparin at the end
of pregnancy can lead to severe haemorrhagic complica-
tions, if premature delivery occurs. The efficacy and safety of
low molecular weight heparin has not yet been assessed
for these patients.

Conclusion
Women who have survived congenital heart disease into
adulthood often have a strong desire to become pregnant.
Optimum care of these potentially complicated pregnan-
cies can only be achieved by a combined approach by car-
diologists and obstetricians in specialist centres with an
understanding of the obstetric and cardiac complications
that can arise.

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2 Siu S, Chitayat D, Webb G. Pregnancy in women with congenital heart
3 Zuber M, Gautschi N, Oechslin E, et al. Outcome of pregnancy in women
4 de Swiet M. Maternal mortality: confidential enquiries into maternal deaths
5 Rosseter JP, Morales AJ, Repke JT et al. A prospective longitudinal evalu-
599–606.
7 Connolly HM, Geoghan M, Barnes CA. Pregnancy among women with con-
genital atrial correction for transposition of the great arteries. J Am Coll Cardiol
8 Neumann U, Somerville J. Outcome of pregnancies in patients with
10 Feldcamp M, Jones KL, Ornony A, et al. Postmarketing surveillance for
angiotensin-converting enzyme inhibitor use during pregnancy—United
with major heart defects: results from first cohort of British collaborative study
12 Botto LD, Mulinare J, Erickson JD. Occurrence of congenital heart defects
recommendations by the American Heart Association. JAMA 1997;277:163.
14 Joffe JA, Page RL. Treatment of cardiac arrhythmias during pregnancy: a