TRANSPOSITION OF THE GREAT ARTERIES WITH ARTERIAL SWITCH OPERATION

Background

Transposition of the great arteries (TGA) describes the anatomic arrangement in which the aorta arises from the right ventricle and the pulmonary artery from the left ventricle. This anatomic arrangement causes blood to circulate in two separate parallel circuits rather than in series.

In approximately 50% of women there are associated lesions, mostly commonly ventricular septal defect and pulmonary stenosis.

Survival without surgery is very poor in newborns born with TGA. The first definitive operations were described by Dr. Mustard and Dr. Senning, but these operations have been abandoned because of the associated late complications. Since the mid 1980s, the operation of choice is the “Arterial Switch Operation”. The Arterial Switch Operation involves transection of the great arteries. The aorta and pulmonary artery are then re-implanted on to the semilunar valves in a hemodynamically “correct” position. The coronary arteries are transferred to the “neo-aorta”. Some women have right ventricle to pulmonary artery conduits depending on the era of repair and associated heart lesions.

Early survival has been reassuring with approximately 95% of women reaching adulthood. Potential late complications include branch pulmonary artery stenosis, neo-aortic valve regurgitation, dilation of the neo-aortic root, supravalvar aortic stenosis, pulmonary stenosis, ventricular dysfunction, and arrhythmias likely related to prior surgeries and coronary obstruction.

This first cohort of women with arterial switch operations are now reaching childbearing age, but experience with pregnancy remains limited.

Effects of Pregnancy-Related Hemodynamic Changes

During pregnancy, cardiac output increases by 50% reaching its peak at around 28-32 weeks of gestation (see Cardiovascular Changes During Pregnancy). The increased cardiac output can result in heart failure in women with impaired ventricular function. Women with stenotic lesions will develop higher gradients in pregnancy. Pregnancy can also precipitate arrhythmia in predisposed women. The hemodynamic and hormonal changes of pregnancy may alter the aortic wall structure and potentially could result in aortic dilation or dissection.

Maternal Complications

Experience in pregnant women with TGA after arterial switch repair is very limited. (1) (2) One small series describes outcomes in 17 pregnancies (4 miscarriage and 13 completed pregnancies). There were no maternal deaths. Two women developed cardiac complications during pregnancy, 1 woman with impaired left ventricular systolic function had non-sustained ventricular tachycardia, and 1 woman with a mechanical systemic atrioventricular valve developed post partum valve thrombosis. With newer modifications of the Arterial Switch Operation, complications are expected to be less common.
Fetal Complications

Data regarding fetal outcomes in this population is limited. In the one small case series, there were no fetal or neonatal deaths in women with arterial switch operations (2). Studies on the entire cohort of women with congenital heart disease have demonstrated an increased frequency of early delivery and smaller infants.

Management Strategies

Preconception Counseling/Contraceptive Methods

Successful pregnancies are reported in women with Arterial Switch Operations; however, systemic ventricular and valve function, aortic root size, and arrhythmia history are important variables to consider when determining an individual’s risk. There are also other cardiac characteristics, which can have an impact on outcomes (see General Considerations).

Ideally, a comprehensive cardiovascular examination should be undertaken before embarking on pregnancy. This includes a careful history and physical examination, electrocardiography and an echocardiogram. The additional prognostic benefit of cardiopulmonary exercise testing has not been defined, but it can useful to assess functional status and ability to increase heart rate during exercise. Diagnosis of coronary artery disease is important; however, the best modality to identify silent ischemia in this population not known. Other imaging modalities such as cardiac magnetic resonance imaging may be useful in select cases e.g. to visualize the pulmonary arteries.

Transmission of congenital heart disease to offspring should be discussed. The risk of transmission of congenital heart disease is approximately 5%, compared to a background risk of approximately 1% of having a baby with congenital heart disease.

A discussion about contraceptive methods is appropriate in all women with TGA. The combined oral contraceptive pill (containing estrogen) is associated with an increased risk of thromboembolism and may be problematic in women with significant ventricular dysfunction, atrial arrhythmias, or mechanical valves. Progesterone-only forms of contraception are not associated with thromboembolic risk and may be suitable alternatives (see Contraception).

Medication use should be reviewed if a woman is contemplating pregnancy or is pregnant. The MOTHERISK website (http://www.motherisk.org) is an excellent resource.

Ante-partum Care

Coordinated care with a congenital heart disease specialist and a high-risk obstetrician should be implemented. The frequency of assessments during pregnancy should be determined on an individual basis.

Because experience with this population is limited, close cardiovascular surveillance is essential throughout pregnancy and in the peripartum period.

Women should be offered a dedicated fetal echocardiogram at approximately 20 weeks gestation.
Labour and Delivery

Labour and delivery should be planned carefully with a multidisciplinary team well in advance. It is important to communicate the delivery plan to the woman and to other physicians involved in her care. The best delivery plan is not useful if information is not readily available when needed.

Generally, vaginal deliveries are recommended unless there are obstetric indications for a cesarean delivery. Good pain management for labour and delivery is very important in order to minimize maternal cardiac stress. To decrease maternal expulsive efforts during the second stage of labour, forceps or vacuum delivery is often utilized. To decrease potential harmful complications from difficult mid cavity-assisted delivery, uterine contractions are often utilized to facilitate the initial descent of the presenting part.

The need for maternal monitoring at the time of labour and delivery is dictated by the women’s clinical status. Most women with TGA do not require invasive monitoring. To detect potential arrhythmias early, continuous monitoring with electrocardiography may be helpful in some instances.

In general, endocarditis prophylaxis at the time of labour and delivery is not recommended in women after arterial switch operations. However, some experts continue to administer antibiotics because they feel that the risks of adverse reactions to antibiotics are small and the risk of developing endocarditis has major health consequences.

Post-partum care

The hemodynamic changes of pregnancy may take up to six months to normalize. Women should be seen early after pregnancy (usually within 6-8 weeks). The frequency of additional follow up visits should be dictated by the clinical status of the women.

References: