TRANSPPOSITION OF THE GREAT ARTERIES WITH MUSTARD OR SENNING OPERATIONS

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. The anatomic arrangement of TGA 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 left ventricular outflow tract obstruction.

Survival without surgery is very poor in newborns born with TGA. The first definitive operations were described by Dr. Mustard and Dr. Senning, hence the names of the respective operations. Both of these operations correct the abnormality by creating a baffle within the atrium in order to redirect (switch) blood flow. This results in repair that leaves the morphologic right ventricle as the systemic (subaortic) ventricle and the tricuspid valve as the systemic (subaortic) atrioventricular valve.

Women born with TGA who have undergone atrial repair by either the Senning or the Mustard procedure have now virtually all reached childbearing age. Potential late complication of the Mustard and Senning operations are tachyarrhythmias (interatrial re-entrant arrhythmias and ventricular tachycardia) and/or bradyarrhythmias (junctional bradyarrhythmia and sinus node dysfunction), systemic ventricular dysfunction, tricuspid (systemic) valve regurgitation, and baffle stenosis/leaks.

Effects of Pregnancy-Related Hemodynamic Changes

After Mustard or Senning operations, the morphologic right ventricle supports the systemic circulation. In many instances there is dysfunction of this subaortic right ventricle. 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 subaortic (systemic) right ventricular function. Dilation of the systemic right ventricle can result in progressive tricuspid regurgitation.

Maternal Complications

According to the Working Group on Pregnancy and Contraception, the overall maternal risk for women with TGA and previous atrial repair is significant (1). However, women with good or only mildly impaired systemic ventricle function, and no history of arrhythmia at the time of conception may have successful pregnancies (2). The most common adverse events during pregnancy are arrhythmias, mainly supraventricular tachyarrhythmia (15.6%) and heart failure (10.8%). (3,4,5,6,7). Thromboembolic events have been described. Maternal deaths have occurred, although rare. There are other cardiac characteristics, which can have an impact on outcomes. (see General Considerations)
Worsening tricuspid regurgitation, right ventricular dilation, and systolic dysfunction are described. In some women, the changes are irreversible. (6) The possible late effects of pregnancy on the heart need to be discussed with women prior to conception.

Women with Mustard or Senning operations are at increased risk for premature labour and hypertensive disorders of pregnancy (7).

Some women with high-risk cardiac lesions may seek alternatives to pregnancy including adoption or surrogate motherhood.

### Fetal Complications

Premature delivery (34%) and small-for-gestational-age babies (19%) are relatively common. Preterm delivery is the leading cause of infant morbidity and mortality in the western world. Fetal and perinatal mortality has been reported to be as high as 4%. (7)

### Management Strategies

#### Preconception Counseling/Contraceptive Methods

Successful pregnancies are reported in women with Mustard and Senning repairs; however, preconception risk stratification is important. Systemic ventricular function, tricuspid valve function, patency of the baffles, and arrhythmia history are important variables to consider when determining an individual’s risk. There are 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, an electrocardiogram 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. Other imaging modalities such as cardiac magnetic resonance imaging may be useful in select cases. Catheterization may be indicated for women requiring surgery prior to pregnancy or if there are other unaddressed hemodynamic issues.

Transmission of congenital heart disease to offspring should be discussed. The risk of transmission of congenital heart disease is approximately 5-10%, compared to a background risk of 1%.

A discussion about contraceptive methods is appropriate in all women with Mustard or Senning operations. Combined estrogen/progestin containing contraceptive should be used with caution, if at all, in women with Mustard or Senning operations and significant ventricular dysfunction or atrial arrhythmias. (see Contraception). Progesterone-only forms of contraception are safe.

Women treated with angiotensin converting enzyme inhibitors or angiotensin receptor blockers will need to have these medications stopped prior to pregnancy. Assessment of ventricular function after discontinuation of therapy is useful. 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. Close cardiovascular surveillance is essential throughout pregnancy and the peripartum period. The frequency of assessments (clinical and echocardiographic) during pregnancy should be determined on an individual basis. Women with good functional capacity, good ventricular function and no cardiac events prior to pregnancy have the best chance for an uncomplicated pregnancy. Nonetheless, these women remain at risk for ventricular dilation and dysfunction and therefore, serial echocardiographic follow up should be performed during pregnancy.

Treatment for symptomatic heart failure may be necessary in some women. The role of beta blockers in women with impaired systemic (subaortic) ventricular systolic function is not known, but they may be helpful.

Supraventricular arrhythmia can be treated medically or with DC cardioversion when women are unstable or unresponsive to medical therapy. (see Arrhythmias)

Volume overload should also be avoided in women with impaired subaortic right ventricular function as it can result in heart failure.

Women should be offered fetal echocardiography 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 the woman’s functional status, the degree of ventricular dysfunction, and associated lesions. 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 Mustard or Senning 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.

Air filters (bubble trap filters) are only recommended in women with TGA after previous Mustard/Senning procedure if there are residual interatrial or interventricular shunts.

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. Because ventricular function may deteriorate after pregnancy, (6) postpartum echocardiographic follow up is important.

References: