

ATRIAL SEPTAL DEFECT

Background

Atrial septal defects (ASD) are among the most common congenital heart lesions. They can occur in isolation or as part of more complex congenital heart defects. This section covers isolated ASD. Atrial septal defects can be classified into 4 types: 1. Secundum type ASD, 2. Sinus venosus defects, 3. Primum ASD (as part of the spectrum of atrioventricular canal defects), and 4. Unroofed coronary sinus ASD. The most common type is the secundum type ASD and this type of ASD is the focus of this section.

Depending on their size, ASD can lead to volume overload of right-sided heart chambers with dilatation of the right ventricle and right atrium. Large ASD are often detected in early childhood and patients undergo surgical or interventional closure. Not uncommonly however, ASD can remain unrecognized until adulthood. ASD may be diagnosed incidentally or may be diagnosed when patients present with complications. Late complications of ASD are right heart failure, atrial arrhythmias (mainly atrial flutter and atrial fibrillation), and paradoxical embolism. Rarely there is an association with pulmonary hypertension and the development of Eisenmenger syndrome (see Eisenmenger Syndrome). The diagnosis of an ASD is sometimes made during pregnancy when an echocardiogram is done to investigate a flow murmur.

Effects of Pregnancy Related Hemodynamic Changes

Pregnancy is associated with hemodynamic changes including an increase in blood volume and cardiac output and a decrease in systemic vascular resistance. These factors can lead to increases in right-sided filling and decreases in left sided pressures. (see Cardiovascular Changes During Pregnancy). These hemodynamic changes are usually well tolerated in women with an isolated ASD or surgically closed ASD, without pulmonary hypertension, with normal right ventricular systolic function, and with no associated defects. Women with more complex ASD can develop arrhythmias or cyanosis.

Women with significant pulmonary hypertension and cyanosis (Eisenmenger syndrome) tolerate the hemodynamic changes in pregnancy poorly and represent a high-risk group. Their management is discussed in the section about Eisenmenger syndrome (see Eisenmenger Syndrome).

Maternal Complications

In women after surgical or interventional closure of an ASD, and those with a small unrepaired ASD, pregnancy is usually well tolerated in the absence of significant right ventricular systolic dysfunction or pulmonary hypertension. Arrhythmias and right heart failure are potential complications occurring in <1% of pregnancies. (3) Women with residual shunts are at risk for paradoxical embolism. Other cardiac characteristics can have an impact on outcomes (see General Considerations).

Fetal Complications

In general, fetal complications are infrequent. The risk of premature delivery and intrauterine growth restriction seems to be comparable to pregnancies in women without heart disease. (1,2,3,4)

Management Strategies

Preconception counseling/Contraceptive methods

Most women with a repaired or unrepaired ASD can have a successful pregnancy. In the absence of ventricular dysfunction or pulmonary hypertension, heart-related complications are rare.

Ideally, a comprehensive cardiovascular examination should be undertaken before embarking on pregnancy. This includes a careful history and physical examination, an echocardiogram and an electrocardiogram. The additional prognostic benefit of cardiopulmonary exercise testing has not been defined.

Indications for ASD closure prior to pregnancy are based on current guidelines. (5) In the absence of standard indications for ASD closure, women should not undergo surgery or intervention for the sole purpose of reducing the maternal cardiac risk during pregnancy.

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 approximately 1% of having a baby with congenital heart disease. However, rarely, ASD may be associated with genetic syndromes such as the Holt-Oram syndrome. Genetic counseling is recommended for women with other congenital defects including non-cardiac defects or those with a family history of ASD or congenital heart disease.

A discussion regarding contraceptive methods is appropriate in all women with ASD. In the presence of an unrepaired or residual ASD, estrogen-containing formulations of contraception are relatively contraindicated due to the risk of paradoxical emboli. In the setting of an ASD with concomitant pulmonary hypertension, estrogen-containing birth control formulations are contraindicated.

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

Ante-partum Care

The place and frequency of antenatal visits with the cardiologist depends on the cardiac status of the women, the degree of right ventricular systolic dysfunction, and if there is any evidence of pulmonary hypertension. Women without the high-risk characteristics do not need to be seen in high-risk specialized centers.

In women with a large unrepaired ASD or high-risk features (significant right ventricular systolic dysfunction or pulmonary hypertension), antenatal care should be provided by a dedicated multidisciplinary team of experienced cardiologists, obstetricians, and anesthetists at a high-risk pregnancy center. In women without high-risk features, antenatal care and delivery can be performed at non-specialized centers.

Women are at risk for paradoxical embolism. Therefore prophylaxis of deep vein thrombosis with low molecular heparin at times of increased risk (i.e. when a patient is bedbound for obstetric reasons) should be considered.

Women should be offered fetal echocardiography at approximately 20 weeks gestation.

Labour and Delivery

In women with high-risk features, 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 as well as other physicians involved in her care. The best delivery plan is not useful if information is not readily available when needed.

Vaginal delivery is preferred. Women with large unrepaired ASD with significant right ventricular systolic dysfunction or pulmonary hypertension (complex ASD) are at higher risk for complications. In women with complex ASD, 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.

Most women with a repaired or unrepaired ASD do not require special monitoring. The need for maternal monitoring is dictated by the functional status of woman, the systolic function of the right ventricle, the degree of pulmonary artery hypertension and the oxygen saturations. In some women at increased risk of arrhythmias, telemetry monitoring may be helpful.

In women with an ASD, air-particulate filters (bubble trap filters) are recommended for intravenous lines to decrease the risk of paradoxical air embolism.

In general, endocarditis prophylaxis at the time of labour and delivery is not recommended in women with ASD.

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.

If an ASD is initially diagnosed during pregnancy, a complete assessment should be performed after delivery.

References:

- Siu SC, Sermer M, Colman JM, Alvarez AN, Mercier LA, Morton BC, Kells CM, Bergin ML, Kiess MC, Marcotte F, Taylor DA, Gordon EP, Spears JC, Tam JW, Amankwah KS, Smallhorn JF, Farine D, Sorensen S; Cardiac Disease in Pregnancy (CARPREG) Investigators. Prospective Multicenter Study of Pregnancy Outcomes in Women With Heart Disease. Circulation.2001;104:515-521.
- Yap SC, Drenthen W, Meijboom FJ, Moons P, Mulder BJ, Vliegen HW, van Dijk AP, Jaddoe VW, Steegers EA, Roos-Hesselink JW, Pieper PG; ZAHARA investigators. Comparison of pregnancy outcomes in women with repaired versus unrepaired atrial septal defect. BJOJ 2009;116(12):1593-601.
- 3. Drenthen W, Pieper PG, Roos-Hesselink JW, van Lottum WA, Voors AA, Mulder BJ, van Dijk AP, Vliegen HW, Yap SC, Moons P, Ebels T, van Veldhuisen DJ; ZAHARA Investigators. Outcome of pregnancy in women with congenital heart disease: a literature review. J Am Coll Cardiol. 2007;49(24):2303-11.

- 4. Zuber M, Gautschi N, Oechslin E, Widmer V, Kiowski W, Jenni R. Outcome of pregnancy in women with congenital shunt lesions. Heart. 1999 Mar;81(3):271-5.
- 5. Silversides CK, Dore A, Poirier N, Taylor D, Harris L, Greutmann M, Benson L, Baumgartner H, Celermajer D, Therrien J. Canadian Cardiovascular Society 2009 Consensus Conference on the management of adults with congenital heart disease: shunt lesions. Can J Cardiol. 2010;26(3):e70-9.