PULMONARY HYPERTENSION

**Background**

Pulmonary hypertension is defined at cardiac catheterization as a mean pulmonary artery pressure of > 25 mmHg at rest or > 30 mmHg with exercise (1). However, the diagnosis is usually suspected following echocardiography. In 2003 the World Health Organization classified pulmonary hypertension into five groups: 1) pulmonary arterial hypertension, 2) pulmonary hypertension with left heart disease, 3) pulmonary hypertension with lung disease and/or hypoxemia, 4) pulmonary hypertension due to thrombotic and/or embolic disease, and 5) miscellaneous group. (1) Pulmonary arterial hypertension can be further classified as follows: 1) idiopathic, 2) associated with collagen vascular disease, congenital systemic to pulmonary shunts, portal hypertension, HIV infection, drugs and toxins, 3) associated with pulmonary venous or capillary disease.

Women with congenital cardiac shunts, significant pulmonary hypertension and cyanosis (Eisenmenger syndrome) tolerate the hemodynamic changes in pregnancy poorly. Their management is discussed elsewhere (see Eisenmenger Syndrome).

Young women with pulmonary artery hypertension embarking on pregnancy are most commonly women with idiopathic pulmonary arterial hypertension or with pulmonary hypertension associated with other disorders (i.e. connective tissue disorders).

**Effects of Pregnancy-Related Hemodynamic Changes**

The hemodynamic changes of pregnancy are usually poorly tolerated in women with pulmonary hypertension (see Cardiovascular Changes During Pregnancy). In women with significant pulmonary hypertension and a low cardiac output state, the compromised right ventricle may not meet the demands of increasing blood volume and cardiac output. In addition, a fixed pulmonary vascular resistance with a resulting inability to increase pulmonary blood flow may not accommodate an increase in cardiac output. Similarly, large fluctuations in blood volume both peri- and post-partum may not be tolerated by an already compromised right heart and pulmonary vasculature.

**Maternal Cardiac Complications**

A systematic review of published studies from 1978-1996 examined maternal mortality in women with primary and secondary pulmonary hypertension and demonstrated mortality rates of 30% and 56% respectively (2). A more recent review suggested that mortality remains high. (3)

The high risk of maternal death is mostly due to the additional cardiovascular stresses that pregnancy places on an overburdened right ventricle. Most complications occur near term and during the first postpartum week, and may include heart failure and sudden death presumably due to arrhythmias. Other clinical symptoms to be aware of include increasing fatigue, worsening peripheral edema, chest pain that could reflect right ventricular ischemia, and presyncope/syncope with exertion reflecting a decrease in cardiac output.
Fetal Complications

Spontaneous abortion is common. Intrauterine growth restriction is seen in 30% of pregnancies as a result of maternal hypoxemia. Premature labour is common, seen in 50-60% of pregnancies, and there is a high perinatal mortality rate due mostly to prematurity (2).

Management Strategies

Preconception Counseling/Contraceptive Methods

Based on the high mortality risk both during pregnancy and peripartum, as well as the suboptimal fetal outcomes, women with pulmonary hypertension should be strongly advised against pregnancy. (2,3). Some women who are fully informed and understand the maternal and fetal risk and complications may still become pregnant, and unfortunately women may present pregnant without having received appropriate preconception counseling.

In the current era, many women with pulmonary hypertension are treated with pulmonary vasodilators. Preconception discussions with a pulmonary hypertension specialist regarding pulmonary vasodilator therapy during pregnancy is important if women are actively trying, against advice, to conceive, as some vasodilators are teratogenic.

The importance of close monitoring with admission to a hospital equipped with a multidisciplinary team experienced in the management of cardiac disease in pregnancy as needed should be conveyed to your patient.

A discussion about contraceptive methods is imperative (4). (see Contraception) Progesterone-only formulations such as depot injections and subdermal implants (Implanon®) are reasonable options. Progesterone-only pills are not optimal because of the unacceptably low efficacy rates. Contraceptive pills containing estrogen (combined contraceptive pills) are contraindicated due to an increased risk of thromboembolism. The insertion of intra-uterine contraceptive device can be associated with vasovagal reactions, which can be devastating in women with pulmonary hypertension. Some women will consider sterilization due to the high-risk nature of a pregnancy. However, such a decision may have a major psychological impact and needs to be fully discussed with the appropriate caregivers. Moreover, the laparoscopic procedure carries risk in this population, as it requires insufflation of the abdomen with carbon dioxide, intermittent head down tilt and positive pressure ventilation, all of which reduce cardiac output and may be poorly tolerated. If sterilization is chosen, it is strongly recommended that it be performed at a centre with experience in the care of patients with pulmonary hypertension/Eisenmenger syndrome. Essure® is a new sterilization technique, involving the insertion of stents hysteroscopically into the Fallopian tubes using sedation and local anesthesia. Early studies suggest it may be safe and have a low failure rate.

As certain anticoagulants and pulmonary vasodilators are contraindicated in pregnancy, medication use should be reviewed if a woman is seriously contemplating pregnancy or is pregnant. The MOTHERISK website is an excellent resource. (http://www.motherisk.org)

Ante-partum Care

If a woman with significant pulmonary hypertension becomes pregnant, coordinated care should be established early, involving a pulmonary hypertension specialist and a high-risk obstetrician at a high-risk pregnancy center. Close cardiovascular monitoring, with specific attention to volume status, is essential throughout pregnancy and the peripartum period.
Management of volume status is imperative. Volume overload should be avoided as it cannot be accommodated by the pulmonary vascular bed and therefore can result in heart failure. Very frequent follow up is necessary during the later stages of pregnancy as cardiac output peaks, and as pulmonary vascular disease may progress. Treatment for heart failure may be necessary and concurrent oxygen therapy may be administered for dyspnea; however, there is no evidence that it improves outcomes. In some instances, bed rest may be considered to reduce cardiac demands.

Early treatment with pulmonary vasodilator therapy should be discussed with a pulmonary hypertension expert. While no pulmonary vasodilators are considered completely safe during pregnancy, there are a number of cases reports/case series in the literature describing the use of inhaled, intravenous and oral pulmonary vasodilators during pregnancy. (5,6,7,8) However, use of bosentan is not advised in pregnancy owing to the teratogenic effects seen in animal studies. (9)

Women with pulmonary hypertension are vulnerable to thromboembolism. Underlying pulmonary thromboembolic disease is common, even in non-pregnant women. Because of this, many women are on anticoagulant therapy and an appropriate anticoagulation plan should be devised with a hematologist/thrombosis expert prior to conception.

Fetal echocardiography can be offered to the expectant mother to screen for congenital heart defects if there is a family history of congenital heart disease. A fetal echocardiogram is done at approximately 20 weeks gestation.

**Labour and Delivery**

Labour and delivery must be planned carefully with a multidisciplinary team well in advance. The plan should be communicated to the patient and tertiary and local healthcare teams.

Successful vaginal and cesarean deliveries have been reported. The decision regarding mode of delivery should be based on the individual patient and the local obstetrical experience. If vaginal delivery is chosen, good pain management is very important. 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.

Maternal monitoring will often include telemetry monitoring, pulse oximetry, and invasive blood pressure monitoring. Invasive pulmonary artery pressure monitoring is not routinely indicated and can be dangerous.

**Post-partum Care**

The mortality risk remains particularly high postpartum and many experts advise an extended prolonged postpartum period of monitoring in hospital.

After discharge, close postpartum monitoring is necessary. Care should be aimed at managing volume status. The risk of pregnancy related complications exists until 6 months post-partum at which time pregnancy related hemodynamic changes will have fully returned to baseline. Follow up at a frequency thought to be appropriate by the physician should take place until 6 months post-partum.

**References:**


