How to manage Pulmonary Arterial Hypertension during pregnancy?

Pulmonary arterial hypertension (PAH) is one form of pulmonary hypertension. This disease is high blood pressure in the pulmonary arteries. It is the pulmonary arteries that carry blood from the heart to lungs. With PAH, the pulmonary arteries abnormally constrict. As a result, the heart is forced to beat faster and causes blood pressure to rise within the lungs. According to the Pulmonary Medicine Journal, PAH in pregnant women carries a high mortality rate from 30 to 56 percent. The physiologic changes that occur during pregnancy and the peripartum period are not tolerated well with women who have PAH. Labor and delivery further increase blood pressure and cardiac output during uterine contractions. The majority of maternal deaths occur within one month postpartum or during labor. Because of the high risk of death of PAH during pregnancy, delivery and the peripartum period, many medical professionals feel that women with PAH should be advised against pregnancy. If a woman with PAH decides to get pregnant, it is imperative that the pregnancy is managed by a multidisciplinary team. The team should include an obstetrician, pulmonary hypertension specialist, neonatologist, cardiologist and an anesthesiologist who is specialized in managing high-risk pregnancies.

Diagnosing PAH in Pregnant Women

Often, fatigue and exertional dyspnea are some of the common symptoms of PAH during pregnancy. Since these same symptoms can occur in a healthy pregnant woman, the diagnosis of PAH may be delayed. Typically, symptoms begin to get worse and exertional dyspnea can happen during rest. Chest pain, ankle edema, elevated jugular venous pressure and a second heart sound are also signs that indicate PAH. Echocardiography and right heart catheterization are used to confirm the diagnosis. Since both of these procedures avoid radiation, there is low fetal risk. It is recommended that pregnant women with PAH stay in a lateral position, reduce cardio output and seek hospitalization in the beginning of the third trimester of pregnancy.

Management and Treatment of PAH During Pregnancy

Currently, there is no standardized approach for the management of PAH during pregnancy. Treatment is developed by a multidisciplinary team for the best outcome. The treatment for PAH complications during pregnancy vary and include inhaled nitric oxide therapy, calcium-channel blockers, sildenafil and targeted pulmonary vasodilators. Nitric oxide therapy has been effective for reducing pulmonary vascular resistance. As a result, the right ventricle is better able to compensate for physiological changes that occur during pregnancy. Calcium-channel blockers are medications that help relax the artery wall muscles. The arteries are widened, and blood pressure is reduced. Diltiazem and nifedipine are two examples of calcium-channel blockers.

If calcium-channel blockers are ineffective or cannot be tolerated, specific target medications like sildenafil and epoprostenol may be used in the treatment plan. Like calcium-channel blockers, sildenafil relaxes the muscles in the walls of the blood vessels and allows them to become more dilated. Pressure in the blood vessels is reduced, and the strain on the heart is reduced. Targeted pulmonary vasodilators include epoprostenol and iloprost. Epoprostenol is also a naturally occurring prostaglandin and a B category pregnancy drug. It is administered intravenously through a narrow tube in the chest or arm. According to the U.S. National Library of Medicine National Institutes of Health, most of the published case reports about epoprostenol involved initiating this drug several weeks before delivery and had a successful maternal outcome. Iloprost has also demonstrated benefits in pregnant women with PAH with no reported mother or infant fatality.

Vaginal Delivery vs Caesarean

There are no case studies that establish whether vaginal delivery or Caesarean is more favorable for the best outcome. The best mode of delivery depends on the individual circumstances and is decided by the multidisciplinary team. Vaginal delivery is associated with volume changes and presents a problem due to limited cardiac output. Pushing may also pose a problem with haemodynamics. Often, Caesarean delivery is done due to premature delivery. A low-dose spinal anesthesia and epidural are preferred over general anesthesia. This method helps avoid vasodilatation. Central venous pressure and haemodynamic monitoring are essential. Hospital observation is often required for at least two weeks following delivery.

In sum, PAH is likely to worsen during labor and delivery and present challenges. Early recognition and treatment by an experienced multidisciplinary team are of high importance for a positive maternal-fetal outcome. It is also important for women with PAH to continue with the recommended therapy postpartum as there have been reports of death in cases where mothers have died when they opted to discontinue therapy after giving birth.