Abstract: Primary pulmonary hypertension is a disease of unknown etiology in which pulmonary artery hypertension exists in the absence of significant parenchymal disease or a primary cardiac disorder. Primary pulmonary hypertension associated with pregnancy carries a high mortality rate of 30-50. Primary pulmonary hypertension is a rare progressive condition aggravated by the physiological changes occurring during pregnancy. Here we describe the case of a twin pregnancy presenting with primary pulmonary hypertension.

Keyword: Pulmonary artery pressure, Primary pulmonary hypertension, twins.

Introduction: Primary Pulmonary Hypertension (PPH) represents Group 1 within the Pulmonary Hypertension (PH) classification system of WHO (Venice 2003 revision) and is one of the five groups. Primary Pulmonary Hypertension (PPH) associated with pregnancy carries a high maternal mortality rate. An early case series reported a 50% mortality rate associated with pregnancy and Primary Pulmonary Hypertension (PPH). A more recent account noted a 30% mortality rate and partly attributed the decline in the mortality rate to earlier recognition, better understanding of the pathophysiology of Primary Pulmonary Hypertension (PPH), along with improvements in medical therapy and critical-care obstetrics. Recognition of the elevated maternal-fetal mortality rate has led physicians to recommend effective contraception and, in the event of a pregnancy, early fetal termination. The maternal mortality rate is related principally to the increased demands on the cardiopulmonary system during pregnancy. Under normal circumstances, increases in cardiac output in the range of 30 to 50%, blood volume in the range of 40 to 50%, and oxygen consumption of 20% are observed during pregnancy. Other physiologic changes include an increase in cardiac output during labour and postpartum intravascular volume shifts resulting from blood loss or diuresis. These physiologic events place a great demand on the cardiovascular system, with the greatest incidence of mortality occurring during the first month postpartum. This is likely related to changes in pulmonary vascular tone due to intravascular volume shifts, hypoxemia, or thromboembolism.

Identification of the hemodynamic changes has led to the use of anticoagulants, oxygen, and vasodilators in the management of these patients. We report a successful maternal-fetal outcome in a pregnant woman in whom Primary Pulmonary Hypertension (PPH) was diagnosed who was treated with vasodilators, anticoagulants, and failure measures and delivered by cesarean section for obstetric indication.

Case study: A 28 years Mrs. X, G2A1, married since 10 years, conceived after infertility treatment presented with increasing dyspnoea, fatigue of several weeks duration, hemoptysis three episodes at 30 weeks of pregnancy. There was no significant past medical history.

On examination she was afebrile, dyspnoea of grade-2, orthopnea(+), mild pallor(+), not icteric, no pedal edema, PulseRate-106/min, Blood Pressure-130/80mmHg, JVP-not elevated. On auscultation she had an accentuated pulmonary component of the 2nd heart sound and a systolic murmur of tricuspid regurgitation. Respiratory system - clear. ECG- sinus tachycardia, right heart strain and 'P' pulmonale. Echocardiogram showed normal intra cardiac anatomy but dilated right atrium and ventricle and mild Tricuspid Regurgitation. Pulmonary artery pressure-119mmHg, Left ventricle function-72%, adequate biventricular function, no pulmonary edema. A mild microcytic anaemia was present.

Identification of the hemodynamic changes has led to the use of anticoagulants, oxygen, and vasodilators in the management of these patients. We report a successful maternal-fetal outcome in a pregnant woman in whom Primary Pulmonary Hypertension (PPH) was diagnosed who was treated with vasodilators, anticoagulants, and failure measures and delivered by cesarean section for obstetric indication.
With low dose dopamine support patient was transferred to intensive care unit. Postoperative echocardiography was unchanged. Antifailure drugs were continued. Intravenous heparin was started 12 hours after surgery. APTT ratio was maintained 2-2.5. One unit of packed cell was transfused(Hb-7.9gms). On the 8th postoperative day Tab.Acitrom 2mg was started. Both babies were given to mother and were discharged after one month. Advised to continue Tab.Acitrom and attend cardiology OPD for further follow up. Presently,she has resumed an active lifestyle as a housewife and mother.

Discussion:
Primary pulmonary hypertension is defined as sustained elevation of pulmonary artery pressure (mean greater than 25mmHg at rest). Pulmonary hypertension is tolerated poorly in the parturient. Deterioration typically occurs in the 2nd trimester and during labour. Women with primary pulmonary hypertension are advised against pregnancy. In early pregnancy, termination is considered. If pregnancy is to be continued, further management will require a multidisciplinary team, delivery planned at 32-34 weeks. Several reports have demonstrated an improvement in haemodynamics and outcome in nonpregnant patients receiving vasodilator and anticoagulation therapy for the treatment of Primary Pulmonary Hypertension (PPH). Subsequently, several reports have described the use of vasodilator therapy during parturition and postpartum with good outcomes. Accordingly, we elected to use sildenafil and heparin for several weeks prior to the expected date of delivery in order to maximize the benefits of this therapy. An important component in the successful management of these patients involves a multidisciplinary team approach with an obstetrician, pulmonary or cardiology specialist, anesthesiologist, and experienced nursing staff.

Conclusion:
In summary, Primary Pulmonary Hypertension (PPH) is likely to worsen during pregnancy and delivery, resulting in a high maternal mortality rate. Early recognition and treatment with vasodilator and anticoagulation therapy may reduce the likelihood of complications. A multidisciplinary team approach to the management of patients with Primary Pulmonary Hypertension (PPH) during pregnancy is of great importance for a successful maternal-fetal outcome. Mode of delivery depends on Obstetric indications.

References:
4. Williams Obstetrics-23rd Edition