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## REVIEW ARTICLE

### PREGNANCY COMPLICATING PATENT DUCTUS ARTERIOSUS WITH EISENMENGER SYNDROME

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#### ABSTRACT

Congenital heart disease (CHD) is associated with significant morbidity and mortality in pregnancy (Whittemore *et al.*, 1982 and Shime *et al.*, 1987). Congenital heart disease in pregnancy is associated with reduced cardiac reserve. A 24 year pregnant female 21 weeks of gestation not diagnosed to have heart disease in the past came with chief complaints of class 3 dyspnoea on exertion. On examination patient had differential cyanosis in the lower limbs, loud pulmonary component of second heart sound and parasternal heave, diagnosed to have Patent Ductus Arteriosus (PDA) with Eisenmenger syndrome on Echocardiography. Laboratory investigation including complete blood picture, renal function and liver function tests were within normal limits. As mortality risk of Eisenmenger syndrome is significantly high in pregnancy, termination of pregnancy was done.

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## INTRODUCTION

PDA is vascular connection between descending thoracic aorta and main pulmonary near the origin of left pulmonary artery. PDA can be complicated by Eisenmenger complex which is associated with shunt reversal and development of pulmonary hypertension. Pregnancy in women with pulmonary hypertension is associated with mortality rate in between 30 % and 56 % (Weiss *et al.*, 1996), pregnancy is poorly tolerated in these patients. Maternal deaths are known to occur either during labour or immediate postpartum period (Smith *et al.*, 2012). Pregnancy with Eisenmenger complex has a maternal mortality rate of 50% and fetal mortality rate of 60 % (Roberts *et al.*, 2003 and Drenthen *et al.*, 2007).

### Case presentation

24 year old pregnant female with period of gestation of 21 weeks presented with history of exercise intolerance, dyspnoea on exertion class 3 (New York heart association). No history of heart disease diagnosed in the childhood. On Examination she was in distress, grade 2 clubbing in both lower limbs and Grade 1 clubbing in both upper limbs.

Her oxygen saturation was 83% in both lower limbs, right upper limb oxygen saturation was 96% and left upper limb was 90% suggestive of differential cyanosis between both upper limbs, peripheral pulses were intact. Cardiovascular system examination showed apical impulse in the 5th intercostal space in the midclavicular line, grade 2 parasternal heave, and loud pulmonary component of second heart sound with no ejection systolic murmur. Respiratory system examination was normal. Her laboratory investigations including biochemistry and complete blood picture were within normal limits. Transthoracic echocardiography showed severe pulmonary artery hypertension, dilated Pulmonary artery, PDA with Bidirectional Shunt suggestive of Eisenmenger syndrome. Patient was started on oxygen therapy. As Eisenmenger syndrome is usually associated with high risk of mortality. Under strict aseptic precautions, proper analgesia, medical termination of pregnancy was done with extra amniotic ethacridine lactate instillation. Patient was given intravenous antibiotic ampicillin 500 mg 3 times daily dose and gentamycin 80mg intravenous twice daily as infective endocarditis prophylaxis and thromboprophylaxis with low molecular weight heparin. Post procedure she was symptomatically better and was counselled about the potential complications of pregnancy in her and advised contraception.

## DISCUSSION

Pregnancy is a state of volume overload due to significant increase plasma volume almost 50% above the baseline during

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third trimester of pregnancy (Hunter and Robson, 1992), which further limits cardiac reserve in the patient patients with congenital heart disease, leading to complications like congestive heart failure and sudden hypoxemic death. Conditions associated with increased sympathetic activity like anaemia, infection, anxiety tilt the compensated heart to state of decompensation. Pulmonary hypertension, Maternal cyanosis, functional status of the mother, maternal anticoagulants predict the outcome of maternal and fetal complications in patients with CHD (Smith, 1998 and Kasper *et al.*, 2005). In general PDA is well tolerated in pregnancy. PDA is functionally closed within 48 hrs of birth, however patency of ductus beyond 3 months is considered pathological. Hemodynamic status of patient in PDA is determined by magnitude of shunting, which in turn depends on flow resistance and pressure gradient across aorta and pulmonary artery. Moderate to large PDA results in pulmonary over circulation and overload on the left side of the heart causing congestive symptoms, although they have minimal symptoms in childhood, many of them become symptomatic at later age especially in third decade of life (Campbell, 1968). Heart in them is associated with atrial fibrillation (Marquis *et al.*, 1982).

Patients with PDA for years develop irreversible pulmonary vascular disease due to progressive increase in pulmonary vascular resistance secondary to long standing pressure overload in pulmonary circulation, although many patients are related to coincident primary pulmonary vascular disease as the pulmonary vascular disease proves to be fatal even after closure of PDA (Espino-Vela *et al.*, 1968 and Bessinger *et al.*, 1975). When the pulmonary vascular resistance increases beyond systemic vascular resistance pulmonary to systemic reversal of shunting and results in development of Eisenmenger syndrome (Wald and Crean, 2010), Once patient develops Eisenmenger syndrome the machinery murmur is no longer heard resulting in difficulty to diagnose PDA. However pathophysiology of Eisenmenger syndrome is not clearly understood. It is said that micro vascular injury stimulates production of growth factors and enzymes which causes intimal proliferation, medial hypertrophy in association with endothelial dysfunction and platelet adhesion leads to obliteration of pulmonary vasculature (Perkett *et al.*, 1990 and Celermajer *et al.*, 2012). Pregnancy should be discouraged in patients with Eisenmenger syndrome as it is associated with significant mortality. High incidence of maternal mortality in Eisenmenger was due to hypovolemia, thromboembolism and Gestational hypertension (Mukhopadhyav *et al.*, 2012). Adoption of advanced therapies and multidisciplinary treatment approach improved fetal and maternal outcome in advanced gestational age group patients with Eisenmenger syndrome where termination is not possible. Patient should be explained about mortality risk in pregnancy and option of surrogacy can be explained.

### Conclusions

Complications of PDA can be prevented by timely arrival of diagnosis and management. Pregnancy is absolutely contraindicated in Eisenmenger syndrome due to significant mortality. Patient counselling to avoid pregnancy in future and Option of surrogacy should be explained.

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