

# Fatal Outcome of Infective Endocarditis in a Pregnant Marfan Woman

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## Abstract

Heart disease complicates 0.5% to 1.5% of pregnancies. Among these, infective endocarditis is rare in pregnancy but can have devastating consequences for both mother and fetus (22.1% and 14.7% mortality rate respectively). Marfan syndrome is one of the most common inherited disorders of connective tissue that can be lethal due to pregnancy related complications. The patient was a 19 year old primigravida at 8 weeks gestation presented with fever since 2 weeks prior to admission. Echocardiography revealed large vegetation on leaflet of the bioprosthetic (porcine) valve which was done previously. Her pregnancy terminated at 12 weeks of gestation spontaneously, but after 8 weeks of antibiotic therapy, due to occurrence of paravalvular leakage, she was taken to the operating room for the second time but couldn't be weaned from cardiac pump and died.

## INTRODUCTION

Heart disease during pregnancy remains the fourth leading cause of maternal death<sup>1</sup>. Its incidence is reported to be between 0.5% and 1.5%<sup>2</sup>. Heart diseases are the most important non-obstetric causes of maternal death during pregnancy, accounting for 10% of maternal deaths<sup>3</sup>. Among cardiac disease, bacterial endocarditis is uncommon during pregnancy (0.006%) but is serious<sup>3</sup>. Despite reported incidence of 1 in 10000 to 20000 individuals of Marfan syndrome in general population<sup>4</sup>, and its known pregnancy-associated complications<sup>5</sup>, endocarditis is not reported during pregnancy. Herein, we report a case of fatal infective endocarditis in a pregnant patient with Marfan syndrome.

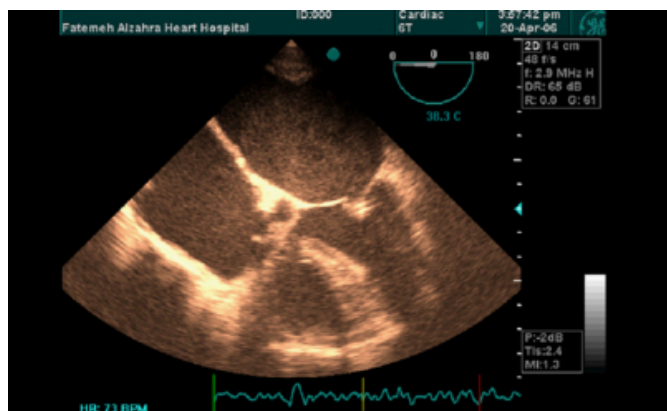
## CASE PRESENTATION

A 19 year old female primigravida at 8 weeks gestation from southern rural regions of Iran who was referred to our center due to fever since 2 weeks prior to admission. She had upper to lower body segment ratio of 0.7, an arm span to height ratio of 1.2, with body temperature of 38.5oc orally. Ophthalmoscopic examination was normal. Arachnodactyly of fingers and toes, with positive thumb and wrist signs were present. An early systolic musical murmur best heard in apex with radiation to her left axilla. Trans esophageal echocardiography showed dilated aortic sinus of Valsalva, mitral valve prolapse and a large (9mm x 10mm) vegetation on mitral valve leaflet leading to moderate mitral regurgitation (Figure). One week later, due to persistence of

fever despite 10 days of optimal antibiotic therapy (ceftriaxone, ampicillin and gentamicin), persistently elevated white blood cell count (18700/c.mm.) ESR (98mm/hr) and CRP (150 mg/liter) and 3 sets of negative blood culture and size of vegetation, the patient was sent for operation. Surgeon found an 8mm x 10mm vegetation so he excised the mitral valve and replaced it with a bioprosthetic (porcine) valve. After operation, the patient did not become afebrile; noteworthy was that all cultures including excised vegetation cultures were negative. Repeated TEE did not reveal vegetation any more. At 12 weeks of gestation, the patient developed vaginal bleeding; sonography showed no fetal heart activity, so dilatation and curettage was done. 1 week later, the patient underwent another TEE that showed 2 small oscillating masses attached to mitral valve leaflets. 2 days later the patient developed sudden onset left upper abdominal pain and tenderness, abdominal CT-scan with contrast showed hypodense area in the periphery of spleen without enhancement by contrast agent suggestive of embolic infarction. After a course of 4 weeks of antibiotic therapy, her fever subsided and antibiotics were continued for another 4 weeks. On termination of antibiotic therapy course, the patient developed sudden onset of dyspnea; TEE examination revealed paravalvular leakage. She was operated again, but could not be weaned from cardiac pump and died.

### Figure 1

Figure: TEE showing vegetation on the mitral valve leaflet



### DISCUSSION AND REVIEW OF LITERATURE

Pregnancy and the peripartum period are associated with important cardio circulatory changes that can lead to marked clinical deterioration in the women with heart disease. Heart disease complicates 0.5% to 1.5% of pregnancies<sup>2</sup>. Among these, bacterial endocarditis is extremely rare, the reported incidence varying widely from 0.003% to 0.013%<sup>7</sup>, in selected reports. The estimated incidence would be 0.005% to 0.015% of pregnancies, as bacterial endocarditis account for approximately 1% of cases of cardiac disease. Its rarity is emphasized by the fact that only 68 authenticated cases of infectious endocarditis complicating pregnancy was found with review of literature from 1965 through 2003<sup>(8)</sup>.

Nazarian et al first described the successful valve replacement in a case of pregnant patient with bacterial endocarditis in 1976<sup>9</sup>. Cox et al describe their experience with seven women whose pregnancies were complicated by endocarditis as streptococcus viridians was the most commonly isolated organism (found in four of seven cases)<sup>10</sup>. In the majority of patients, the disease has tended to run a subacute course, with the offending organism being streptococcus viridians. Acute bacterial endocarditis, on the other hand, is most commonly caused by staphylococcus aureus.

Marfan syndrome one of the most common inherited disorders of connective tissue, is an autosomal dominant condition with a reported incidence of 1 in 10000 to 20000 individuals<sup>4</sup>. Although there is a wide range of clinical severity associated with Marfan syndrome there are certain features involving the skeletal, cardiovascular, and ocular involvement. Aortic root disease, leading to aneurysmal dilatation, aortic regurgitation, and dissection, is the major cause of morbidity in the Marfan syndrome<sup>4</sup>. Despite a lot of

studies of pregnancy in Marfan syndrome<sup>5,11,12,13</sup>, there is no report of endocarditis in pregnancy, just there is a case of endocarditis after delivery of a Marfan patient<sup>14</sup>.

To our best knowledge this case is the first reported case of infective endocarditis during pregnancy in a Marfan patient.

### CONCLUSION

Infective endocarditis is rare in pregnancy but can have devastating consequences for both mother and fetus. Marfan syndrome is one of the most common inherited disorders of connective tissue that can be lethal due to pregnancy-related complications. It seems that association of both diseases in a pregnant woman is highly lethal.

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