

Multidisciplinary Management Of Marfan Syndrome in Pregnancy: A Case Report

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Introduction

Marfan syndrome (MS) is a multisystem autosomal dominant connective tissue disorder with a prevalence of 1 in 5000. Common manifestations are aortic root dilatation, lumbar sacral ectasia and scoliosis. We present a case demonstrating the need for multidisciplinary input and imaging to provide optimal care.

Case Report

35yr primigravida presented in pregnancy with MS, with a positive fibrillin 1 gene. Her mother died of complications of MS at 24yr postpartum. Our patient had a pre pregnancy cardiology assessment and echo review, which demonstrated an aortic root 41mm. She was advised to proceed with aortic root surgery and mitral valve repair pre pregnancy. Surgery was complicated, requiring further bypass and coronary artery stenting. She also sought genetic counselling and had in vitro fertilisation with pre implant genetic diagnosis. Antenatal assessment included spinal MRI, which demonstrated lumbar dural ectasia, expansion of the perineurium, scalloping of lumbar sacral bony anatomy and significant anterior meningocele, considered a low pressure cyst.

Continue Case Report

Neurosurgery advised delivery by Caesarean due to abnormal anatomy, which would likely delay normal progression of labour and also advised against regional anaesthesia. Cardiology assessment in early pregnancy with echo demonstrated moderate aortic regurgitation and normal systolic function. Blood pressure was controlled on bisoprolol. The anaesthetic plan was to proceed under general anaesthesia with intra-arterial monitoring. Opioids were administered on induction to obtund pressor response and videoscope to facilitate intubation due to a high arch palate. Partial view of the vocal cords was visualised on laryngoscopy. Phenylephrine was the vasopressor of choice. Surgery was uneventful with our patient discharged on day 2.

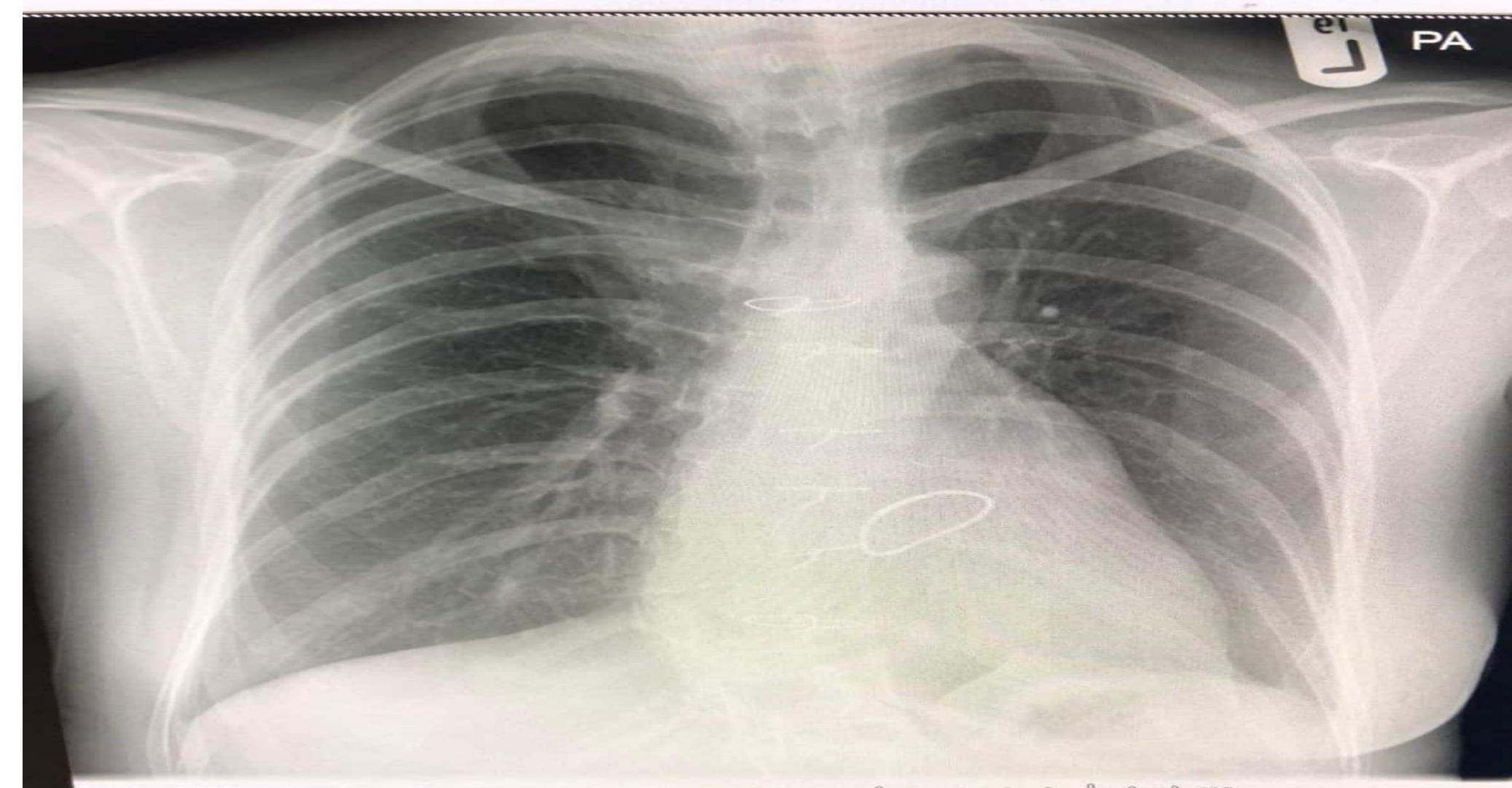


Figure 1 : CXRay showing thoracic scoliosis , metallic mitral valve & sternotomy scar

Discussion

There are no clear guidelines on the anaesthetic management of women presenting in pregnancy with MS. Epidural, spinal and combined spinal-epidural have all been administered with variable success. Ideally, patients present for pre pregnancy counselling and assessment. We advise multidisciplinary review with the use of imaging to determine a safe and successful outcome for each individual patient with MS.

References

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