A Multisystem Disorder with Multisystem Challenges: Perioperative Management of a Parturient with Marfan’s Syndrome, Severe Scoliosis, and High Arching Palate

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Marfan’s syndrome is a systemic connective tissue disorder caused by a genetic mutation on chromosome 15, resulting in an abnormal extracellular matrix protein. It demonstrates an autosomal dominant inheritance pattern, and the worldwide incidence is approximately 1 in 5,000.1 Marfan’s syndrome is associated with joint laxity and scoliosis. In addition, Marfan’s patients often demonstrate serious cardiac abnormalities including mitral valve prolapse and ascending aortic dilation or dissection. Pregnancy related cardiovascular changes have the potential to create catastrophic outcomes for both the mother and the fetus in the setting of Marfan’s syndrome. Research also suggests that pregnancy is associated with a high rate of premature deliveries, preterm premature rupture of membranes and increased mortality in the offspring.2 We present a case of successful anesthetic management of a Cesarean delivery in a patient with Marfan’s syndrome and severe scoliosis.

A 25 year old Gravida 1, Para 0 at 38 weeks gestational age, with Marfan’s syndrome and severe scoliosis, presented for Cesarean section. The patient was eleven years status post aortic root and aortic valve replacement with recent cardiac MRI demonstrating normal LV and RV size and function, appropriately seated prosthetic aortic valve and root, and no mitral valve prolapse. Additionally, she had a history of scoliosis status post Harrington rod placement and bilateral ectopic lentis. Her exam was remarkable for a high arching palate and a MP class III airway. Her spinous processes were palpable despite the severe curvature of her spine. Anticoagulation was maintained with a heparin drip, which was discontinued and PTT normalized prior to surgery. An arterial line was placed in addition to IV access prior to anesthetic intervention. A spinal was attempted without success, and the decision was made to convert to general anesthesia. She was induced with propofol and succinylcholine and rapidly intubated using a glide scope. The patient and her baby boy did well post-operatively; however, on POD#2, after re-initiation of anticoagulation, her course was complicated by abdominal bleeding requiring multiple transfusions and exploratory laparotomy.

Pregnancy is associated with increased risk of aortic dissection in Marfan’s patients, probably caused by hemodynamic changes and hormonally mediated loss of elastic fibers in the aortic wall.3 Pre-operative management must include a thorough history and physical exam along with cardiac evaluation throughout pregnancy in association with a cardiologist.