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Perioperative Medical Management of the Marfan Patient Undergoing Repeat Cardiothoracic Surgery

Aashish Shah, MD, and Adam Skrzynski, MD
Oakland University, William Beaumont School of Medicine, Royal Oak, MI

Marfan syndrome is an autosomal-dominant connective tissue disease that impacts multiple organ systems (including the cardiovascular system), various tissue properties, bone calcification, and pulmonary parenchyma. Common cardiothoracic surgical procedures in this population include repair of the aorta, cardiac valves, and coronary arteries. These patients undergo multiple procedures leading to scarring and poor wound healing, which further complicates medical management. Often, these surgeries are emergent, without any opportunity for perioperative medical clearance.

Case Presentation: A 30-year-old man with a history of Marfan syndrome presents to the hospital with a 40-pound weight loss over 9 months and complaints of shortness of breath on exertion. He is found to have a subtherapeutic international normalized ratio. His past medical history is significant for type A aortic dissection requiring aortic valve replacement with a St. Jude mechanical valve and mitral valve prolapse. On physical examination, the patient's appearance was remarkable for marfanoid body habitus; fingers demonstrated arachnodactyly; chest examination revealed pectus carinatum; and there was extreme laxity of joints in all limbs. A grade 5/6 systolic ejection murmur was auscultated at left sternal border.

Initial management included intravenous heparin drip, serial electrocardiograms, transthoracic echocardiogram and transesophageal echocardiogram. The patient was found to have a flailing mitral valve leaflet. After expert cardiology and cardiothoracic surgery consultations, the patient underwent valve repair surgery and was placed on cardiopulmonary bypass. After an unsuccessful attempt at repair, a St. Jude mechanical valve was placed in the mitral position with an intra-aortic balloon pump inserted to ensure adequate cardiac output.

The patient's perioperative and postoperative courses were complicated. Immediately following surgery, the patient went into atrial fibrillation and was found to be in cardiogenic shock with an ejection fraction of 20%. Four days after surgery, he started to experience altered mental status and left arm weakness. Radiographic imaging of the brain revealed a right-sided infarction. Other complications included a right groin hematoma at the surgical catheterization site, persistent hyponatremia, and poor oral intake requiring insertion of a feeding tube. After a prolonged period of mechanical lung ventilation, a tracheostomy was required. Once the patient stabilized, he was transferred to an inpatient rehabilitation unit. More than 1 month following his admission, he was finally discharged home.

Conclusion: Appropriate perioperative medical clearance is vital in all patients undergoing cardiothoracic surgery, especially those predisposed to anatomic and physiologic aberrancies such as Marfan patients. This case highlights the need for appropriate perioperative evaluation in patients with Marfan syndrome undergoing repeat cardiothoracic surgery.