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A case of Marfan syndrome with thoracic aortic root dilatation

Stanlies D'Souza

Baystate Health, dsouzastan@yahoo.com

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Introduction

This case involves the anesthetic management of a 15-year-old patient with thoracic aortic root dilatation presenting for multi-level spinal fusion for idiopathic scoliosis. The importance of perioperative hemodynamic monitoring and smooth arterial pressure management is emphasized in order to prevent aortic dissection.

Case Description

A patient with thoracic aortic root dilatation in Marfan Syndrome has an increased risk of perioperative aortic dissection, making strict control of blood pressure (BP) essential in the perioperative period. A 15-year-old, 92 kg male with Marfan Syndrome with thoracic aortic root dilatation presented for T3-L4 posterior spine instrumentation and fusion (PSIF). Preoperatively, he was started on losartan 50 mg, which was continued in the postoperative period. An arterial line was placed for close monitoring of BP in the intraoperative and postoperative period. We tightly controlled patient's BP during induction, intubation, intraoperative period, extubation and postoperatively.

Case Discussion

Cardiovascular manifestations of Marfan Syndrome¹

- ✓ **Thoracic aortic root dilatation: Usually asymptomatic, frequency 60-80%, aortic dissection is the most common complication. Dissection is rare in children under the age of 10.**
- ✓ **Pulmonary artery dilatation: Usually asymptomatic, frequency 75%, dissection is rare**
- ✓ **Mitral valve prolapse, mitral regurgitation: Palpitations are usual symptoms, frequency 50-70%, arrhythmias may be present**
- ✓ **Descending aorta dilatation: Usually asymptomatic, frequency 80-100%, increased risk of dissection in adults**
- ✓ **Tricuspid valve prolapse: Patients most commonly asymptomatic, incidence is around 4%**
- ✓ **Left ventricular dysfunction: Common presenting symptom is dyspnea. Frequency is 100%, initially diastolic and may progress to systolic dysfunction.**

Medical therapy to prevent Progression of Aortic Root dilatation and Aortic Dissection¹

1. Beta blockers are the main mode of therapy²
2. Calcium channel blockers
3. Angiotensin converting enzyme inhibitors
4. Angiotensin receptor blockers

Indication for Aortic Root Replacement Surgery in Thoracic Aortic Root Dilatation³

1. Thoracic root diameter > 5 cm
2. Progressive dilatation > 1 cm/year
3. Progressive aortic regurgitation

Management Strategy for Patients with Marfan Syndrome with Thoracic Root Dilatation presenting for Major Surgery

1. Close hemodynamic monitoring with invasive arterial line
2. Avoid acute hemodynamic changes
3. Continue the elective antihypertensive therapy in the perioperative period

CONCLUSION

In our case of multi-level posterior spinal fusion for idiopathic scoliosis, we continued losartan in the perioperative period and avoided swings in blood pressure during critical times in the perioperative period.