Anesthetic Management of a Patient with Williams Syndrome and Failure to Thrive for Reimplantation of an Aberrant Right Subclavian Artery Requiring One-lung Ventilation

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INTRODUCTION

Described in 1961 by Williams et al, Williams Syndrome (WS) is a rare genetic disorder caused by a deletion of 26 contiguous genes on chromosome 7, most notably the elastin gene. The absence of elastin in vascular wall construction results in widespread arterial disease including supravalvular aortic stenosis, supravalvular pulmonic stenosis, coronary disease and diffuse aortic stenosis. These cardiovascular abnormalities are the primary concern when providing anesthesia and sedation to patients with WS. Multiple cases of cardiac arrest and sudden cardiac death have been described. We present the successful anesthetic management of a patient with WS who underwent reimplantation of an aberrant right subclavian artery using left thoracotomy and one-lung ventilation.

CASE DESCRIPTION

We present a 2 year-old female with WS and failure to thrive found to be secondary to an aberrant right subclavian artery causing esophageal compression. Surgery was planned for reimplantation of the artery via a left thoracotomy to relieve the compression. Prior to induction all records including imaging, previous anesthesia records, and a transthoracic echocardiogram were reviewed. The patient was induced using a 50:50 mixture of oxygen and nitrous oxide with sevoflurane carefully titrated to hemodynamics. Upon confirmation of induction a 22 ga IV was placed in the left hand and nitrous oxide was discontinued. Endotracheal intubation was facilitated with fentanyl (2 mcg/kg) and vecuronium (0.1 mg/kg). Using the Storz C-MAC to visualize the glottis a 5 Fr bronchial blocker and 4.0 mm micro cuff endotracheal tube were placed through the vocal cords.

Figure 1) Barium swallow study performed due to failure to thrive showing esophageal compression found to be caused by an aberrant right subclavian artery

DISCUSSION

The patient was turned left lateral and a caudal block was performed using 12 mL of 0.1% ropivacaine and 250 mcg of preservative-free morphine (25 mcg/kg) for postoperative analgesia. The patient was turned back supine, a 20 ga IV was placed in the right foot, and a 22 ga left radial arterial line was placed. Final bronchial blocker placement and lung isolation was confirmed with fiberoptic bronchoscopy (FB). The patient was positioned in the right lateral position for surgery. Bronchial blocker position was again confirmed with FB and lung isolation initiated. Anesthesia was maintained with sevoflurane, vecuronium, and fentanyl, while the aberrant right subclavian artery was successfully reimplanted and esophageal compression relieved. Dexmedetomidine infusion was started after reimplantation and the patient was extubated and transferred to the ICU without incident and without evidence of discomfort.

CONCLUSION

The cardiovascular abnormalities associated with WS pose a significant risk for patients undergoing anesthesia. Unfortunately, this risk is compounded by the fact that anesthesia and/or sedation is frequently required to facilitate characterizing these anomalies. Even patients who have previous uneventful anesthesia, have a risk of a cardiac event with subsequent anesthetics. Thus, appropriate preparation and careful management are critical to provide safe anesthesia. By utilizing a neuraxial block we were able to decrease the incidence of bleeding for a major vascular surgery and the dose of all maintenance anesthetics thereby decreasing myocardial depression and cardiac events.

REFERENCES

Matisoff et al., Risk assessment and anesthetic management of patients with Williams syndrome: a comprehensive review. Paediatr Anaesth. 2015 Dec;25(12):1207-15