Williams Syndrome: A Day in the Dental Room

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Introduction

Some of the most difficult patients to manage as pediatric anesthesiologists are children with Williams syndrome (WS). The incidence of WS is approximately 1 in 10,000 - 20,000 live births. Their underlying genetic abnormality is a deletion on chromosome 7q11.23 which leads to a defect in the elastin gene and subsequently the elastic structure of vascular walls. The most concerning aspect of their care is the risk of peri-procedural cardiac arrest.

In general, children with WS have identifiable facial features, developmental delay, failure to thrive as infants and significant cardiac disease. Due to their multisystemic illness, they tend to have several procedures requiring either sedation or general anesthesia in their lifetime. We will discuss the anesthetic management of a patient with WS undergoing dental extraction.

Case Report

A 7yo 23.7 kg M with WS, severe supravalvar aortic stenosis (SVAS) (Figure 1), left ventricular hypertrophy, stenotic RCA, dilated LAD, severe coarctation, and asthma presents for dental extraction prior to cardiac surgery.

He was pre-medicated with 2mg intravenous (IV) midazolam and taken to the OR. After application of ASA monitors, the patient underwent controlled induction with lidocaine, additional midazolam, titrated fentanyl, etomidate, rocuronium, 1% sevoflurane and intermittent phenylephrine boluses to maintain pre-operative blood pressures. He was then orally intubated and a radial arterial line was placed. Due to ST changes with minimal hypotension while using low inhaled anesthetic doses, the patient was started on a low dose vasopressin infusion to maintain mean arterial pressures within 10 percent of baseline. During the dental extraction, he had multiple episodes of tachycardia with associated hypotension which required treatment with esmolol. He was reversed with a titrated glycopyrrolate dose to prevent excessive tachycardia and extubated deep on dexmedetomidine. He was then admitted to ICU for monitoring while awaiting his cardiac surgery.

Anesthetic Considerations: Non - Cardiac

Neuro:

- Severe developmental delay, highly social/enthusiastic personality but with significant anxiety and phobias - may have difficulty with mask induction and could benefit from **premedication** and/or child life

- Hypotonia- problems with coordination

Facial Features:

- Wide mouth, broad forehead, short nose with broad tip, mandibular hypoplasia - may be a **difficult intubation**

Endocrine:

- Commonly have hypercalcemia as well as impaired glucose tolerance

Gastrointestinal:

- GERD

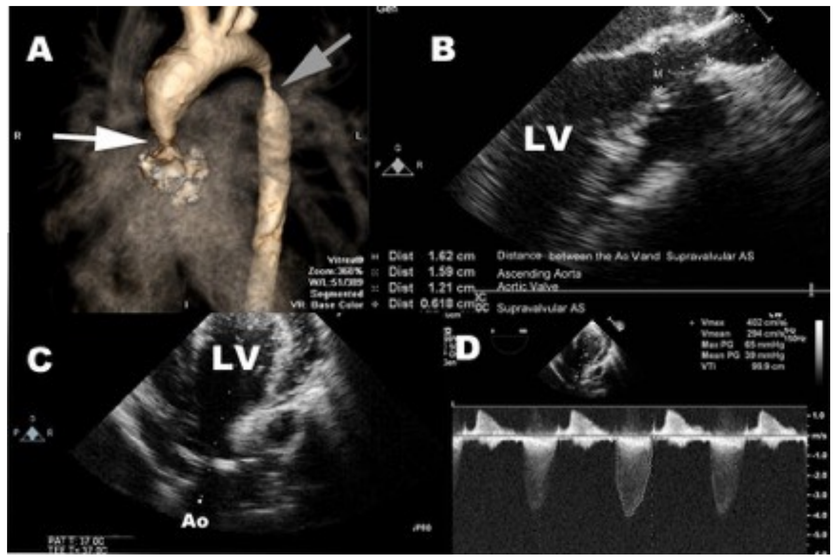

FIGURE 1:

A) CT angiogram with 3D reconstruction exhibiting a supravalvular aortic stenosis of 8 x 8 mm (white arrow) and discrete coarctation of 6 mm distal to the left subclavian; B) TEE mid-esophageal aortic long axis view showing the supravalvular stenosis at 16 mm of the aortic valve (12 mm) and the post-stenotic aortic dilation (15.9 mm); C) TEE deep transgastric view displaying the alignment of the Doppler with the LVOT and ascending aorta; D) Continuous wave Doppler at the same level showing flow acceleration with a mean gradient of 39 mmHg and peak gradient of 65 mmHg.

Anesthetic Considerations: Cardiac

Supravalvar aortic stenosis:
- usually at level of sinotubular junction
- develop compensatory LVH which leads to increase O2 demand

Supravalvar pulmonary stenosis:
- most commonly seen as peripheral pulmonary artery stenosis
- frequently seen in combination with SVAS

Coronary anomalies:
- most commonly seen as left coronary stenosis or ostial slit
- may occur with or without SVAS
- increased risk for coronary ischemia due to anomalies and increased demand

Arrhythmias:
- prolonged QT which may worsen with tachycardia
- need to be treated promptly and aggressively to prevent further increase in demand and worsening ischemia

Generalized management of cardiac concerns

- Maintenance of pre-operative blood pressure within 10% of baseline
- Maintenance of normal sinus rhythm
- Maintenance of pre-load and SVR
  - can be achieved through: maintenance fluids (especially if patient is not the first case and is NPO) and vasopressors (neosynephrine/vasopressin)
- Avoidance of tachycardia
  - can be achieved through: appropriate analgesia/anxiolytics
- Prevention of elevated pulmonary vascular resistance
  - can be achieved through: elevated FiO2, hyperventilation, normothermia, avoiding acidosis, preventing sympathetic surges

- Emergencies:
  - Consider having ECMO back up available as patients can decompensate quickly and be difficult to resuscitate

Discussion

As a pediatric anesthesiologist, you are more likely to see a patient with Williams Syndrome in the general OR going for one of the many surgeries that they may require throughout their lifetimes. They may present with supravalvar aortic stenosis, supravalvar pulmonary stenosis, peripheral pulmonary artery stenosis, coronary anomalies, prolonged QT, developmental delay and difficult airways. We should always consider invasive monitoring, be prepared for cardiac arrest, and the need for emergent circulatory support. With the possibility of rapid and difficult to treat cardiac decompensation, ECMO can provide life saving temporization if resuscitation efforts are unable to reverse ischemia and improve cardiac function. Due to the potential risk of severe complications, these patients should to be done in a tertiary center and never as outpatients. Intra-operative goals include maintenance of pre-operative blood pressures, normal sinus rhythm, avoiding tachycardia, maintaining preload and systemic vascular resistance, and prevention of elevated pulmonary vascular resistance.

REFERENCES

1. Collins RT, et al. Peri-procedural risk stratification and management of patients with Williams syndrome. Congenital Heart Disease. 2017;12:133–142.

2. Burch, T. M., et al. (2008). Congenital supravalvular aortic stenosis and sudden death associated with anesthesia: What's the mystery? Anesthesia & Analgesia, 107(6), 1848-1854.

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