Anaesthetic challenges in peri-operative management of williams syndrome

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ABSTRACT

Introduction: Williams syndrome is a rare microdeletion disorder associated with significant craniofacial and cardiac anomalies. The unique presentation of this syndrome and increased risk for sudden cardiac death poses a significant challenge to the anaesthesiologist to cater to varied requirements in airway and intraoperative management. The cardiac anomalies associated with this syndrome are complex and require prompt surgical correction.
Case report: A 2 and half year old male child, a known case of william's syndrome underwent surgery for correction of supravalvular aortic stenosis. He succumbed to sudden cardiac arrest in the immediate postop period. Discussion: Williams syndrome are associated with multifaceted cardiac defects with poor prognosis due to increased risk of sudden cardiac death and cardiac arrest resistant to normal resuscitation. This emphasises the need for careful and thorough perioperative care taking it to consideration the unique problems associated with this syndrome. Conclusion: A case of willams syndrome who underwent cardiac surgery is presented here with special emphasis on the challenges and distinct issues faced in the perioperative period
Key words: Williams syndrome, Cardiac arrest, Cardiac surgery, Anaesthesia

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INTRODUCTION

William syndrome, is a rare microdeletion disorder initially described in 1961 by Williams, Barratt-Boyes, and Lowe.[1] It is characterised by developmental abnormalities, craniofacial features and cardiac anomalies. It has an incidence of 1 in 10,000 live-births. [2] A triad of supravalvular aortic stenosis, mental retardation and elfin facies characterize this disease. Anaesthetic management of these patients is a risky proposition because of difficult airway due to micrognathia and mandibular retrusion and a higher predilection for sudden cardiac arrests thus necessitating greater vigilance during anaesthesia and the post-operative period. Anaesthetic management of Williams syndrome [WS] for cardiac surgery is discussed here.

CASE REPORT

A two and a half year old child a known case of Williams syndrome with supravalvular aortic stenosis [SVAS], pulmonary artery stenosis and severe mitral regurgitation was admitted for correction of SVAS. The child had delayed developmental milestones and typical elfin facies.

The hematologic and biochemical parameters were within normal limits. Chest X ray showed cardiac silhouette consistent with biventricular hypertrophy. Echocardiography revealed supraaortic long segment, ascending aortic stenosis with gradient of 65mmHg, severe mitral regurgitation, calcified mitral leaflet, branch pulmonary artery stenosis and a large left atrium. Cardiac catheterization confirmed the diagnosis and the coronary vessels were normal.

On the operation day, the child was premedicated with ketamine 7mg/kg and midazolam 0.2 mg/kg intranasally. Once the child was sedated, a 22G peripheral intravenous line was inserted. Child was then induced with Fentanyl 10mcg/kg and rocuronium 1mg/kg. Trachea was nasally intubated with 5.0mmID uncuffed endotracheal tube. Maintenance was achieved with sevofluorane 1 MAC in a 1:1 oxygen:air mixture. After induction, femoral artery and femoral vein were cannulated. Right subclavian vein was cannulated for measuring left atrial pressure post-operatively. He was monitored intra-operatively for peripheral arterial oxygen saturation [SpO2], electrocardiography, rectal and nasopharyngeal temperature, urine output, intra-arterial blood pressure, central venous pressure, left atrial pressure,and end-tidal carbon dioxide.

After median sternotomy, pericardial patch was harvested. Child was heparinised and an activated coagulation time >480seconds was achieved before he was was put on cardio-pulmonary bypass using aortic and bicaval cannulation. Heart was arrested with antegrade cardioplegia. Left atrium was opened and mitral valve was assessed. Annuloplasty was done with a strip of bovine pericardium. Aorta was opened and enlarged with pericardial patch. Right pulmonary artery ostium was enlarged with pericardial patch. The surgical procedure progressed uneventfully without significant hemodynamic or technical problems. Once the child was rewarmed and correction of routine physiological and biochemical parameters were achieved the aortic cross clamp was removed and child was successfully weaned off cardiopulmonary bypass[CPB]. CPB time was 187 minutes and aortic cross clamp time was 124 minutes. Trans-esophageal Echocardiography [TEE] gradient across aortic valve was corrected from 60 mmHg to 15 mmHg. After surgery, the child was shifted to ICU with vasopressor support

Two hours after surgery, the child suffered a sudden cardiac arrest with ventricular fibrillation and hypotension. Immediate cardio-pulmonary resuscitation with external cardiac massage and inotrope was initiated but the child was in continuous ventricular fibrillation. Emergency sternotomy with internal cardiac massage and defibrillation were attempted. He was revived but thereafter was in a semi-comatose state with neurological obtundation and ventilator dependence. Tracheostomy was done for airway protection. After 2 months, he succumbed to illness.

DISCUSSION

Williams syndrome is a rare congenital anomaly which poses a great challenge to the anaesthesiologists. Cardiac anomalies in Williams syndrome include supravalvular aortic stenosis, narrowing of peripheral systemic and pulmonary arteries. Supravalvular aortic stenosis is a complex and uncommon disease that is treated by surgical intervention. Despite

meticulous care sudden cardiac arrest and death have been reported during anesthesia, surgery and invasive procedures like cardiac catheterization.[3-6] Many of these patients have not responded to standard cardiopulmonary resuscitation. The distinct character of cardiac deaths in this patient is that there is rapid, sudden deterioration which is refractory to resuscitation. The risk-benefit ratio should be worked out before subjecting a patient with willams syndrome to any diagnostic test. The parents should be involved in the decision making process and should be thoroughly explained the potential consequence of the test or procedure.

Bird et al [3] noted that the crucial points where maximum death occurred were either immediately at the start of the procedure or post . He pointed out that the severity of SVAS is not a predictor of sudden death which was confirmed by Wessel et al also.[8] The presence of coronary artery stenosis and severe biventricular outflow tract obstruction was implicated as two most important anatomical abnormality associated with sudden cardiac death. The plausible mechanisms of death for these anatomic abnormalities in these patients are myocardial ischemia, decreased cardiac output, and arrhythmia. Developing preventive strategies based on these observations, to formulate monitoring techniques to identify this group of patients with increased risk of sudden cardiac death will be the logical next step in management. This was evident from our case, which had a sudden cardiac arrest in the ICU which was refractory to conventional resuscitation measures and responded only to internal cardiac massage.

A careful preoperative evaluation, including cardiac catheterization, electrocardiogram, and echocardiogram and proper selection of surgical treatment method are imperative for successful management of these patients.

The anaesthetic goal in these cases are to maintain normal heart rate and rhythm , optimize intravascular volume status, avoid fluctuation in systemic and pulmonary vascular resistance and avoid agents that cause myocardial depression.[9]

Sudden hemodynamic variation leads to a sudden drop in cardiac output and compromises the coronary flow which is further detrimental to the inherent coronary artery obstruction and ventricular hypertrophy. These factors may cause acute decompensation leading to sudden cardiac death.. Although epinephrine may be life-saving, avoidance of multiple or large doses of epinephrine should be considered as this has the potential to increase systemic vascular resistance which in turn can decrease coronary blood flow in the setting of SVAS. Systemic blood flow should be maintained with effective chest compressions rather than the repeated administration of epinephrine. In our case all attempts at vasopressor and inotropic support failed including external chest compression till the time internal cardiac massage was instituted.
The craniofacial features may pose a challenge to anaesthesiologist during tracheal intubation as elucidated by Medley et al [10] who reported a case of difficult intubation with smaller endotracheal tube than expected for age. The characteristic facies which included a flattened mid-face, wide mouth and poor dentition pose a significant difficulty to mask ventilation and tracheal intubation.[10]

Drugs should be used in small incremental doses without causing haemodynamic stability at all times. Intensive monitoring and early recognition of signs of hemodynamic deterioration is warranted to avoid any mishap. Elective procedures should be performed in institutions capable of providing ECMO in the event that conventional resuscitation fail. [12]

CONCLUSION

William syndrome offers a multitude of issues which is of concern to the anaesthesiologist. The different cardiac abnormalities and varied presentation and risk need to be considered while evaluating the patient for surgery. Although all patients with WS need early and ongoing cardiovascular evaluation and follow-up, most patients will not require cardiovascular interventions. [12] Careful selection of cases and vigilant assessment and monitoring is key to prevent adverse events and successful management.

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