Case Report

Anesthetic Management of a Patient with First Arch Syndrome and Acute Valvular Infective Endocarditis Presenting for Full Mouth Rehabilitation

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Abstract

Clinical effects of multivalvular disease is complex making anesthetic management of noncardiac surgery a challenge. Generalizations can be made about hemodynamic goals, but pathophysiology is dynamic and unique to each patient and must be treated as such. The combination of aortic regurgitation (AR) and mitral regurgitation (MR) is usually poorly tolerated because the double-valve regurgitation tends to exacerbate left ventricular volume overload. When these cases are complicated with craniofacial abnormalities, overall anesthetic challenge is amplified.

Keywords: Acute aortic valve insufficiency; Acute mitral valve insufficiency; Difficult airway; Endocarditis; First arch syndrome

Introduction

Patients with acute multivalvular regurgitations are critically ill and hemodynamically compromised. One cause of non-ischemic valvular regurgitation is infective endocarditis (IE). Left-sided, double valve IE is rare but not unheard of [1]. In patients with native valve disease, the prevalence of multiple valve involvement has been reported as high as 20.2% [2]. Urgent/emergent surgical treatment is often required either via valve repair or valve replacement [3,4]. Prior to valve replacement, perioperative dental care is commonly requested in an attempt to minimize the risk for late development of prosthetic valve endocarditis.

The anesthetic management for non-cardiac surgery in this patient population is complex. The risk of major adverse event associated with dental extraction prior to cardiac surgery is as high as 8% [5]. Pre-existing co-morbidities further complicate an already challenging anesthetic particularly when a difficult airway is anticipated. Despite the prevalence of multivalvular disease and its hemodynamic impact on anesthetic management, there is a paucity of discussion in literature to guide perioperative care. We aim to contribute to this body of work by presenting our unusual patient.

Case description

A 22-year-old Caucasian male presented to the operating room for full mouth rehabilitation in preparation for early simultaneous aortic and mitral valve replacement. Past medical history included first arch syndrome, cognitive delay, attention deficit and hyperactivity disorder and hyperthyroidism. Home medications included clonidine, divalproex, olanzapine, pantoprazole and ondansetron.

He was transferred to our facility in severe sepsis and diagnosed with bacterial endocarditis. He was admitted to the ICU with initiation of antibiotics. Physical exam was notable for tachypnea, tachycardia, poor dentition, petechial rash, splinter hemorrhages, and arthritis along with a grade III murmur heard best at lower left sternal border. Relevant lab test results included platelet count of 82,000/mL and acute renal insufficiency (BUN of 43mg x dL\(^{-1}\) and creatinine of 1.5mg x dL\(^{-1}\)).

Over the course of 4 days before the dental procedure, serial echocardiography and CT scans showed progressive worsening of endocarditis and bilateral pleural effusion. Two-days prior to dental restoration, TTE exam found normal left ventricular size and systolic function with estimated LV ejection fraction of 40%, estimated RV systolic pressure of 40mmHg, severe mitral regurgitation from a flail posterior leaflet (Figure 1), and large...
circumferential pericardial effusion with early tamponade physiology (Figure 2). A pericardial drain evacuated 800mL of serosanguineous fluid that day upon insertion. The night before surgery, the pericardial drain put out another 1000mL. He was admitted to the intensive care unit (ICU) with on-and-off norepinephrine infusion prior to coming to the operating room.

Figure 1: Transthoracic echo parasternal long axis view of the left ventricular showing a posterior mitral leaflet flail segment (blue arrow) and a large pericardial effusion (red arrow).

Figure 2: Transthoracic parasternal short axis view of the left ventricle showing larger pericardial effusion (blue arrows).
Preoperatively, the patient was assessed in the ICU where he had been awake and alert and did not complain of distress; however, it was apparent that he is dyspneic when speaking. According to his mother, he received sedation on multiple occasions for dental restoration, but as far as they know, he had not received general anesthesia previously. Mother recalled past anesthetic providers voicing concerns about his airway. His parents, who claimed no family history of anesthetic complications, provided his history.

On airway exam, it was clear that he suffered from craniofacial anomalies. Patient presented with bilateral micrognathia, maxillary hypoplasia and external ear deformities as seen in Figure 3. His mouth opening was limited without ability to protrude his mandible. His palate was high-arched with a Mallampati class III airway. There was no limitation on neck range of motion.

![Figure 3: AP and lateral images of a patient with First Arch Syndrome showing micrognathia, maxillary hypoplasia and external ear deformities along with high-arched palate.](image)

After informed consent was obtained, the patient was transported to the operating room (OR). Standard ASA monitors were applied. Bilateral chest tubes were placed on suction. Patient had an in situ 20-gauge radial arterial line. Using aerosolized 4% lidocaine, patient’s airway was anesthetized. Then, he was carefully sedated with midazolam, fentanyl, and ketamine in small incremental doses and fiber optically intubated through the mouth using a size 7.5 endotracheal tube over a pediatric flexible bronchoscope. Intubation proceeded with ease. His airway was secured on the first attempt with stable vital signs throughout procedure (heart rate of 143 beats per minute, blood pressure of 130/55mmHg and oxygen saturation in the low 90%). Total sedative doses for intubation included midazolam 1mg, fentanyl 25mcg, and ketamine 20mg.

However, after intubation, it quickly became apparent that patient did not tolerate 0.4% expiratory level of sevoflurane and positive pressure ventilation. Heart rate stayed in the 130’s to 140’s, but the systolic, diastolic, mean arterial blood pressures and pulse pressure began to fluctuate wildly beat-to-beat. Some blood pressure numbers registered include 109/10mmHg and 55/26mmHg all of which pointed to impending hemodynamic collapse. Phenylephrine 50mcg was bloused, sevoflurane discontinued, and the patient was able to breath spontaneously after which, he recovered immediately. It was decided that anesthesia will be maintained with a low-dose propofol infusion at 25mcg x kg x min-1 with carefully titrated boluses of midazolam and ketamine for the remainder of the case. The Dentists were able to supplement analgesia with local anesthetics. Hemodynamics were very stable for the remaining operative experience.

At the end of the dental extractions, patient was intubated and transported in stable condition back to the ICU. On the day following dental procedure, TTE exam showed additional severe aortic regurgitation with vegetation (Figure 4), posterior pericardial effusion with fibrinous strands throughout and an echogenic mass within the pericardium likely representing a new vegetation. Five days later he underwent successful aortic and mitral valve replacement.
Discussion

Our patient presented with multiple active physiologic derangements. He suffered from sepsis, severe acute aortic and mitral valve regurgitation, pericardial and bilateral pleural effusions all of which interacted to produce complex and sometimes conflicting hemodynamic goals. His physiology was dynamic, changing from moment to moment, and difficult to predict. His cardiac output and propensity for forward flow is also likely to be exquisitely sensitive to changes in preload, afterload, contractility and other variables like intrathoracic pressure. The fact that he was alive and appeared to be in no distress, despite critical illness, speaks to the vitality of youth and the aegis of prior good health and we were wary to disrupt his delicately compensated physiology.

Ideally, perioperative management would be guided by echocardiography. Direct visualization of the cardiac chambers negates any guesswork on diagnosing the cause(s) of hypotension. Unfortunately, transesophageal echocardiography (TEE) hinders dental extraction, and transthoracic echocardiography (TTE) was unavailable to us during the case.

First arch syndrome, which was first described in 1889, occurs roughly in 1:50,000 live births. The most prominent abnormalities are malar and maxillary hypoplasia, cleft palate, and the absence of the zygomatic arch. Since it is a developmental defect, the presentation and airway implication are on a spectrum [6,7]. Examples of first arch syndrome include Treacher Collins and Pierre Robin syndrome — classic causes of difficult intubation. Since our patient had never been intubated and was unlikely to tolerate prolonged hypoxemia/hypercarbia, we decided to proceed with fiberoptic intubation.

We were uncertain how this patient’s physiology would respond to some events not uncommonly anticipated with awake fiberoptic intubation — coughing, Valsalva maneuver, hypoxia and hypercarbia (and their effects on pulmonary vascular resistance and catecholamine release), and the need for positive pressure ventilation (PPV). While PPV can be lifesaving, it certainly has impact on left ventricular (LV) and right ventricular (RV) physiology that may or may not be favorable depending on other parameters [8]. For these reasons, we aimed to secure the airway as smoothly and comfortably as possible. We chose to sedate our patient, dosing in small mindful increments, since patient and family gave clear history of having tolerated sedation in the past for dental procedures.

As our case illustrates, pre-induction arterial line promotes rapid recognition and intervention when hemodynamic decompensation is imminent. On a beat-to-beat basis, arterial waveform graphically illustrates the dynamic interplay between the stroke volume, the speed of the volume ejected, the distensibility of vascular tree, and the flow rate of ejected blood from central arterial compartment to the periphery [9].

Although generalizations can be made about perioperative management goals, we cannot overemphasize the importance of treating each patient’s pathophysiology as unique and dynamic, one that can shift in an instant. For example, a common recommendation for management of mitral regurgitation is to keep heart rate in the high normal range (80 to 100 beats per minute), [10] a good starting point, yet our patient had been in sinus tachycardia in the

Figure 4: Transthoracic echo parasternal long axis view of the left ventricle showing a posterior mitral leaflet flail segment (blue arrow) and now a large vegetation of the aortic valve (red arrow).

Conclusion

Anesthetic management for non-cardiac surgery of patient with acute multivalvular regurgitation, sepsis and pericardial effusion is challenging particularly when echocardiography is unavailable or infeasible. Hemodynamic goals may be conflicting, dynamic and hard to predict. The impact of positive pressure ventilation and effects of difficult airway further contribute to complexity. Vigilance and thorough understanding of pathophysiology and pharmacology helps supports individualized care of these critically ill patients through the perioperative period.

References