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Anesthesia for a Patient with Uncorrected Tetralogy of Fallot Undergoing a Gynecological Procedure: a Case Report

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Abstract:
Patients with uncorrected Tetralogy of Fallot (ToF) that survive to middle age are very rare. It has been reported that around 10% of affected persons can survive to adulthood, and only 5% reach 40 years of age. Cardiovascular defects lead to compensatory mechanisms that pose various hemodynamic shifts, hence a challenge in administering anesthesia for this limited population undergoing any surgical procedure.
Presentation: A 33 year old female, known case of cyanotic congenital heart disease (ToF), pulmonary tuberculosis category I (ongoing intensive phase treatment), presented with a large abdominopelvic mass. The patient underwent a gynecologic procedure under combined thoracic epidural and general anesthesia with intraoperative use of invasive arterial monitoring.
Intervention: Perioperative diagnostic tools such as echocardiography, 12-lead electrocardiography, and blood tests were utilized to guide perioperative planning. General anesthesia with low dose sevoflurane, thoracic epidural anesthesia, dobutamine and norepinephrine infusions, and goal-directed fluid therapy using arterial pulse pressure variation were utilized.
Outcomes: The patient tolerated the surgery well under combined general and epidural anesthesia. However, in addition to the patient's baseline condition, the absence of an intensive care unit admission and close monitoring led to the patient's demise thirteen hours postoperatively.
Unique learning point and contribution: Postoperative care and monitoring is equally as important as preoperative and intraoperative care and management. A case of an uncorrected ToF undergoing a gynecologic procedure pursued as life-saving, is high risk for postoperative complications, not even a good candidate for surgery altogether. Hence, close monitoring and documentation in an intensive care unit (ICU) setting is deemed absolute for any surgical and anesthetic complications.

Keywords: Tetralogy of Fallot, General Anesthesia, Epidural Anesthesia, Gynecological Procedure

Introduction:

Tetralogy of Fallot is one of the commonest forms of cyanotic congenital heart disease, accounting for 7–10% of all congenital cardiac malformations with an incidence of one in 3,500 live births (1). It is characterized by four cardinal features: ventricular septal defect (VSD); right ventricular (RV) outflow tract obstruction, which is often dynamic; an overriding aorta; and right ventricle hypertrophy (RVH). The degree of RV outflow tract obstruction, the relative pressures in the right and left ventricles, and the proportion of the aorta overriding the VSD determine the presentation and severity of this condition. The

anesthetist can encounter ToF in a variety of settings, ranging from the acutely cyanotic infant to an adult with previous ToF repair presenting for unrelated surgery.

Betranou, et al., reported survival among unoperated ToF patients to be 66% at 1 year of age, 40% at 3 years, 11% at 20 years, 6% at 30 years, and 3% at 40 years (2). Determining at what age a patient's surgical risk outweighs the benefits of operating is controversial and must be made in the context of the patient's clinical circumstances and personal desires. Anesthetic management on these patients can be challenging and is associated with high mortality hence ideal invasive monitoring for real-time assessment of cardiac output, stroke volume, and systemic vascular resistance is vital.

Case Description:

A 33 year old female, known case of cyanotic congenital heart disease (uncorrected ToF), pulmonary tuberculosis category I (ongoing intensive phase treatment), presented with a large abdominopelvic mass. Findings were suggestive of bilateral ovarian new growth probably malignant with complications.

On admission, the patient exhibited generalized weakness with anorexia, difficulty of breathing, and severe abdominal pain. The patient was bedridden and could hardly communicate. Echocardiogram showed concentric remodeling with adequate wall motion and contractility and preserved ejection fraction (54%), dilated right sided chambers with signs of volume and pressure overload, moderate to severe tricuspid regurgitation, mild aortic regurgitation, pulmonic regurgitation, possible infective endocarditis with an impression of Congenital heart disease, Tetralogy of Fallot (Overriding aorta, large perimembranous ventricular septal defect, right ventricular hypertrophy, to consioder subpulmonic pulmonic stenosis), to consider Tricuspid valve endocarditis. Chest radiograph showed findings suggestive of pulmonary tuberculosis with cicatricial changes and granuloma formation; right-sided aortic arch. Hemoglobin, hematocrit and coagulation parameters were elevated with the following values: Hgb 180, Hct 0.57, PT 17.2 (12.6), PTT 42.1 (30.38); electrolytes were within normal limits as seen in Table 1. The patient was co-managed by multiple services such as Obstetrics and Gynecology, General Medicine, Hematology, Anesthesiology, Nursing Service, Cardiology, Infectious Diseases, Supportive, Hospice and Palliative Medicine; and a multidisciplinary conference (MDC) was held to discuss treatment and patient options, attended by all services. The complexity of the case and the extreme risk for perioperative mortality was explained at length to the patient and her family. It was emphasized that perioperative mortality is very possible as the patient was borderline heart failure and the stress of surgery together with induction of anesthesia will likely lead to full blown heart failure. It was also stressed out that the patient was not a good candidate for surgery. Despite these concerns, the surgery proceeded as per patient's request from her desire to be relieved of the massive discomfort she was experiencing from the mass. The MDC was concluded with plans such as the surgery should be fastest as possible to achieve some relief of pain, co-stabilization with pressors, epidural anesthesia for possible intraoperative and postoperative use, and absolute intensive care unit admission. The plan given by Anesthesiology was clear to all services. The use of volatile anesthetics were contingent on patient's tolerance of general anesthesia. The thoracic epidural was for possible intraoperative use but was placed definitely for modulation of postoperative pain. The surgical plan was exploratory laparotomy, peritoneal fluid cytology, unilateral salpingo-oophorectomy, and palpation of nodes.

Pre-operatively, the patient was started on Dobutamine infusion at 5 mcg/kg/min and Norepinephrine at 0.1 mcg/kg/min, with a target mean arterial pressure (MAP) of more than 65mmHg. On induction, vital signs were BP 88/56, HR 96 bpm, RR 22 cpm, O2 75% at 10 LPM via face mask. The patient was placed

on left lateral decubitus position, an epidural catheter was inserted at level T8. An arterial line was then inserted at the left brachial artery, and a right internal jugular catheter inserted, size French 7. Midazolam 5mg and Fentanyl 50mg intravenously were given at this time. Rapid sequence induction was then started using Fentanyl 300mcg, Rocuronium 50mg using video laryngoscopy with an endotracheal tube size 7.0, secured at level 18 and was maintained with Sevoflurane 1-2% volume. The surgery lasted for an hour with no significant intraoperative events, patient stable and tolerated the procedure well. Norepinephrine was downtitrated to 0.05 mcg/kg/min; patient was off pressors prior to transout with the following vital signs: BP 90/60, HR 100s bpm, O2 75% hooked to a mechanical ventilator with the following settings: ACVC, TV 280, RR 14, BUR 14, PEEP 2, Fi02 30%. At this point, it was communicated to the primary service that an ICU admission is a must, however failed to secure one hence the patient was transferred to a regular ward.

Postoperatively, the patient was monitored in a regular ward, not in an intensive care unit which was previously agreed upon in the multidisciplinary conference (MDC). Four hours postoperatively, BP was noted to be 80/47 hence Norepinephrine 0.1 mcg/kg/min and Dobutamine 5 mcg/kg/min were restarted. Six hours postoperatively, hypotensive episodes recurred at 80/50, accompanied by tachycardia at 120s bpm. Norepinephrine was uptitrated to 0.3 mcg/kg/min and Dobutamine 5 mcg/kg/min maintained. Point of care testing was done which revealed the following values: pH 7.326, pCO2 46, PO2 34.5, BE(ecf) -2, BE(b) -2.4, spO2 61.3%, Na 140, K 3.5, Ca 1.14, Cl 106, Hct 51, Hgb 17.3, Glu 142, Lac 0.83, BUN 16, Crea 0.84, as seen in Table 2. Eight hours postoperatively, still with hypotensive episodes and tachycardic, Norepinephrine was uptitrated to 0.4 mcg/kg/min, Dobutamine 5 mcg/kg/min maintained, Vasopressin 0.03 iu/min started. Twelve hours postoperatively, persistent tachycardia at 180-200s bpm, SBP 80-90 were observed. Continuous decline in vital signs eventually led to a code and advanced cardiovascular life support (ACLS) was done with 7 cycles of resuscitation. There was no return of spontaneous circulation and time of death was announced thirteen hours postoperatively.

Discussion:

Anesthetic management of a patient with uncorrected ToF for noncardiac surgery is a challenge to anesthesiologists and requires a thorough understanding of pathophysiology, events and effects of medications which can alter the magnitude of right to left shunting (3). Goals of anesthesia are to avoid changes in systemic vascular resistance (SVR) to prevent altering any existing shunt since increase in SVR decreases right to left shunting and improves oxygenation; Avoid increases in pulmonary vascular resistance (PVR) by preventing hypoxia, hypercarbia, hypothermia, acidosis, and providing supplemental oxygen that leads to increased right to left shunting; Maintain normal to elevated cardiac filling pressures especially in patients with right ventricular impairment; Continuous ECG monitoring due to the high incidence of both atrial and ventricular arrhythmias; Tachycardia and increases in myocardial contractility should be avoided; Avoid hypothermia as this may increase PVR (4).

Premedication with anxiolytics and hypnotics must be undertaken very cautiously because hypoventilation and hypercapnia may produce deleterious increases in PVR, particularly in patients with underlying pulmonary hypertension or systemic to pulmonary shunts (5). However, patients with chronic hypoxemia retain a normal ventilatory response to hypercarbia as well as to opioid analgesics (5).

Patients with poor cardiac function, who require inotropes preoperatively, may not tolerate inhalational induction, and favor the use of ketamine (6). Ketamine is the preferred induction agent in ToF due to its property of increasing SVR which decreases right to left shunting (7). At lower concentrations,

administration of volatile anesthetics improves arterial oxygenation by causing relaxation of the muscle spasm in RVOT and decreases the total body oxygen consumption (8). Inotropes should be continued and intravenous induction agents titrated (6). The need for invasive monitoring should depend on the type of surgery and cardiac lesion (9). Management of pain is an important concern in these patients because increased sympathetic activity due to pain may trigger cyanotic spell in the perioperative period (3). Development of cyanotic spell can happen due to spasm of hypertrophied pulmonary infundibulum. Tachycardia and increased myocardial contractility can cause this and must be given thorough consideration. It responds to volume, increase in SVR with alpha agonists such as Phenylephrine or Ephedrine (10). Pain management is a critical factor during intra and postoperative management. Opioid infusion for major operations has been the primary postoperative intervention for pain for these patients (11,12). The use of regional anesthesia for well-compensated patients was reported with no complications (13).

The major risks during the postoperative period include bleeding, dysrhythmias, and thromboembolic events (5). Patients presenting with severe congenital heart disease and/or high risk surgery should be managed if possible in a postoperative intensive care unit experienced with caring for adults with congenital heart disease.

Conclusion:

Carefully administered anesthesia with all hemodynamic goals in mind, especially in patients with uncorrected ToF, combined with thorough and conscientious planning, rational drug use, strict close monitoring, vigilance and mindful execution can deliver a safe outcome even in the most difficult of cases.

Appendix:

Table 1. Laboratory Results

CBC	
Hgb	180
Hct	0.57
WBC	7.0
Plt	339
Chemistry	
Na	136
K	4.5
Ca	2.03
Crea	45
PT/PTT	
PT	12.6/ 17.2
INR	1.39
PTT	30.38/42.1

Table 2. Point of care testing values

pH	7.326
pCO2 mmHg	46

pO2 mmHg	34.5
HCO3 mmol/L	24
BE(ecf) mmol/L	-2
BE(b)	-2.4
SpO2%	61.3
Na mmol/L	140
K mmol/L	3.5
Ca mmol/L	1.14
Cl mmol/L	106
Hct	51
Hgb g/dL	17.3
Glu mg/dL	142
Lac mmol/L	0.83
BUN mg/dL	16
Crea mg/dL	0.84

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