A 24-year-old woman with a history of unrepaired tetralogy of Fallot was scheduled to undergo laparoscopic cholecystectomy. Her significant history included tetralogy of Fallot with pulmonary atresia, hypoplastic left pulmonary artery, pulmonary vascular obstructive disease, a functioning right subclavian artery to right pulmonary artery shunt (modified Blalock-Taussig palliative procedure) with a similar shunt on the left side that is occluded.

The patient underwent general endotracheal anesthesia for laparoscopic cholecystectomy for cholelithiasis and pancreatitis. Anesthetic induction, intraoperative course, and the postoperative period proceeded uneventfully, and the patient quickly progressed to the preoperative level of functioning. The careful application of pharmacological and physiological principles guided the anesthetic plan and produced a successful outcome. Principles for the anesthetic management of the patient with cyanotic congenital heart disease undergoing noncardiac surgery are reviewed.

Key words: Congenital heart disease, laparoscopic cholecystectomy, tetralogy of Fallot.

Introduction

Fallot’s tetralogy consists of a large, unrestrictive, ventricular septal defect, a dilated aorta that overrides the ventricular septal defect, and a right ventricular outflow tract obstruction (pulmonary artery stenosis), which is valvular, infundibular, or both. The last feature of the tetralogy is right ventricular hypertrophy, due to the pressure of the right ventricle pumping against the high pressures of the aorta. The resulting right-to-left shunt increases circulating desaturated blood and gives rise to persistent arterial desaturation and cyanosis. The amount of shunt produced is governed by the right ventricular outflow tract obstruction (which is relatively fixed), the systemic vascular resistance (SVR), and the pulmonary vascular resistance.

The long-term effects of congenital heart disease continue as the child gets older, either before repair or in the unusual case of a patient who has not undergone repair. Compensatory polycythemia increases with time and results in an increased incidence of pulmonary, renal, and thrombotic events. Cerebral abscesses are life-threatening events that may occur as a result of bacterial seeding into areas of previous thrombotic infarction. Aortic valve insufficiency results in chronic biventricular failure and hypoxia. Cardiomyopathy arising from right ventricular hypertrophy and failure is the usual cause of death in adulthood (Figure). Hemoglobin values within the normal range represent a relative anemia in these patients. Blood flow is preferentially shunted to the heart, brain,
and kidney with decreased flow to skin, muscle, bone, and splanchnic circulation, resulting in decreased somatic growth and increased metabolic rate.\textsuperscript{1,6}

The most dangerous consequence of this pathophysiology is a hypercyanotic episode, or "tet" spell, that results from a sudden decrease in SVR or a sudden increase in oxygen consumption.\textsuperscript{4} These attacks occur in 20\% to 70\% of children with tetralogy of Fallot.\textsuperscript{1} Traditional treatments are alpha agonists and fluids to increase SVR and \beta-adrenergic blocking agents to decrease infundibular spasm. Intraoperative tet spells can be broken by deepening the level of anesthesia with a volatile agent. Oxygen and morphine sulfate are also useful adjuncts to therapy. Morphine provides a negative inotropic effect on the infundibulum and may have central nervous system effects to help break the cycle of hypoxia and agitation.\textsuperscript{8,9}

Corrective procedures are palliative or definitive. Palliative procedures include balloon dilation performed during cardiac catheterization or various types of systemic pulmonary artery shunts designed to increase pulmonary blood flow.\textsuperscript{10} The modification of the Blalock-Taussig shunt, consisting of a Gore-tex graft placed between the subclavian artery and the ipsilateral pulmonary artery, is the preferred palliative procedure due to the lesser incidence of pulmonary artery distortion in these shunts.\textsuperscript{11} The aim of shunt placement is to provide increased pulmonary blood flow and allow for growth of frequently diminutive pulmonary arteries in preparation for complete repair. Some patients may have such diminutive or hypoplastic pulmonary arteries that complete repair cannot be obtained.\textsuperscript{12} However, most experts believe that early complete repair should always be the goal, and any increased risk is justified due to the avoidance of palliative procedures (which must be reversed at complete repair) and the chronic consequences of hypoxemia.\textsuperscript{1,13}

Even in the patient who has undergone definitive repair, residual defects occur due to the magnitude of the lesion corrected and its close proximity to the conduction system of the heart.\textsuperscript{14} These defects include pulmonary hypertension, residual ventricular septal defect, tricuspid, aortic, or pulmonary regurgitation, right ventricular dysfunction, right ventricular outflow tract obstruction, right bundle branch block, and ventricular dysrhythmias.\textsuperscript{10,15,16} Even asymptomatic patients who have undergone surgical repair have working capacities, cardiac outputs, and maximal heart rates that are less than normal.\textsuperscript{1,17}

Case summary

\textit{Preoperative period.} A 24-year-old, 54-kg woman was seen in the emergency room and diagnosed with cholelithiasis and cholecystitis. Her medical history was significant for unrepaired tetralogy of Fallot with pulmonary atresia. The physical examination revealed a II/VI systolic murmur and a diastolic murmur. An electrocardiogram (ECG) demonstrated sinus rhythm with right ventricular enlargement, right atrial enlargement, and left atrial enlargement. The atrial enlargements were indicative of a long-standing pathophysiologic condition and resulted from the requirement of increased atrial pressure that had to be generated to bypass the pulmonary artery stenosis. The cardiac catheterization report revealed pressures as follows: right atrium, 11/5; right ventricle, 172/14; left ventricle, 126/16; cardiac index, 3.6; Qp/Qs, 0.9; SVR, 26 Wood's units; and pulmonary vascular resistance (PVR), 2.3 Wood's units.

The elevated right atrium pressure is transmitted pressure from the chronically elevated right ventricle pressure that equalizes across the ventricular septal defect. This left ventricle pressure, although within normal limits by physiologic norms, is elevated in this patient with a congenitally small,
hypoplastic left ventricle. The Qp/Qs reflects the right-to-left shunt, and the increased SVR is probably to compensate for the elevated PVR. A quantitative ejection fraction was not done during this study due to technical difficulty. Because of the equilibration of pressures across the right and left ventricles, univentricular failure rapidly becomes biventricular. A pulmonary flow scan revealed that 43% of perfusion bypassed the lungs totally, with poor perfusion of the left lung, indicative of the occluded Blalock-Taussig shunt.

The chest x-ray revealed mildly increased pulmonary vascularity with rib notching noted, suggesting systemic to pulmonary collateral vessels. This is a common finding in patients with tetralogy of Fallot, as the body attempts to increase pulmonary blood flow via collaterals. The patient’s serum electrolytes and coagulation studies were normal, and liver function test results were elevated due to biliary obstruction. The patient's baseline arterial blood gas results revealed a pH of 7.44; Paco2, 37.8; PaO2, 43; HCO3, 25.5; base excess, 2.2; and an oxygen saturation of 80%. The cardiology consultant recommended subbacterial endocarditis prophylaxis and maintenance of SVR in the perioperative period.

- **Perioperative period.** The patient was prepared for surgery with standard peripheral arterial, and central line access, with midazolam in 1- to 2-mg boluses for anxiolysis. Baseline hemodynamic values were heart rate, 90; blood pressure, 120/72 mmHg; and oxygen saturation, 82%. The ECG showed sinus rhythm.

Intravenous induction was accomplished with etomidate, 18 mg, in divided doses and ketamine, 20 mg. Rocuronium was used for intubation, using rapid-sequence techniques due to compromised oxygenation and decreased functional residual capacity. Rocuronium, fentanyl, and isoflurane 0.7% with 100% oxygen at 2 L/min flow was used for maintenance of anesthesia.

After intra-abdominal insufflation, hemodynamic measurements remained essentially unchanged, with peak airway pressures reaching 38 cm H2O. The ECG leads in II and V5 continued to show sinus rhythm without evidence of ischemia.

Peak airway pressures were at 30 cm H2O at the time of gallbladder removal, and the hemodynamic values remained unchanged. The patient spontaneously recovered neuromuscular function by the end of the case. After assessment of respiratory drive and muscle tone, she was suctioned and extubated in the usual manner. Total crystalloid replacement was 1,200 mL of lactated Ringer’s solution. Urine output for the 1-hour, 35-minute case was 350 mL.

- **Postoperative period.** The patient was transported safely to the postanesthesia care unit, where the ECG rhythm and hemodynamic value remained stable. Arterial blood gases revealed a pH of 7.29; PaO2, 45; PaCO2, 46; base excess, -3.5; and oxygen saturation of 74% on a trial of room air. The patient was maintained on oxygen via nasal cannula for 4 hours postoperatively. The patient remained overnight in the hospital and was discharged the following day. No anesthetic or surgical complications occurred. The patient was called at home 4 days postoperatively, and no problems with anesthesia or surgery were identified.

**Discussion**

A neonate with congenital heart disease is born every 15 minutes in the United States. Improved morbidity and mortality in the presence of associated anomalies have succeeded in making it more common for these children and adults to undergo cardiac and noncardiac surgery. While patients who have not undergone repair are more commonly encountered in larger medical centers, physiologic repairs have residual effects that the anesthetist must be cognizant of when formulating an anesthetic plan.

Infective endocarditis is a major concern for all patients with congenital heart disease. Sterile technique in line insertion and prophylaxis for subbacterial endocarditis with antibiotics is indicated. This is also true for patients who have undergone palliative or corrective repairs. Air bubbles are a preventable cause of perioperative morbidity, especially in patients with shunting, as air or particulate matter may be shunted directly into the arterial tree. Ideally, the anesthetic should result in the maintenance and improvement of the PVR/SVR ratio in cyanotic shunt lesions, such as tetralogy of Fallot. The SVR must be maintained or even increased, while the PVR is decreased to increase pulmonary blood flow and function of the right side of the heart. Because most intraoperative manipulations increase PVR (such as sympathetic stimulation and encroachments on lung volumes), ventilatory control is crucial. Since such control is under the disposition of the anesthetist, it can be manipulated by the use of 100% oxygen and hypocapnia to decrease PVR. The SVR should be aggressively maintained by the use of pressor agents, such as phenylephrine, if needed. Increases in SVR decrease right-to-left shunting and improve arterial oxygen saturation.

Alterations in pharmacokinetics and pharmacodynamics occur in the patient with tetralogy of Fallot, as with any cyanotic heart lesion with right-to-left shunting. Theoretically, the speed of intravenous induction in these patients would be short-
ened, because systemic venous blood bypasses the pulmonary circuit and, therefore, appears in arterial blood quicker. Inhalational agent uptake would be slower due to the diminished pulmonary blood flow, which is only partially relieved by the Blalock-Taussig shunt. Altered uptake and elimination were not clinically appreciable in this specific case, supporting literature that states that these differences are not clinically significant.11,22

The use of nitrous oxide as an adjunct to anesthesia is controversial in these patients. Nitrous oxide is thought to increase PVR in adults but not in children; any intravascular air bubbles will expand in the presence of nitrous oxide and, therefore, increase the deleterious effects of air emboli.4,9 The conduct of the anesthetic should be tailored to the individual patient's cardiovascular and physiologic status. Because of the variability inherent in these patients, there is no consensus in the literature on which agents are best.8,21

Anesthesia for the adult with tetralogy of Fallot requires careful preparation of the operating room. Cardiac support drugs should be immediately available and the anesthesia machine carefully checked preoperatively because of the low margin for error in these patients. Adequate oxygenation is essential to the survival and well-being of these patients in surgical settings.11,12

Monitoring needs for cyanotic patients undergoing noncardiac surgery include ECG, precordial or esophageal stethoscope, noninvasive blood pressure monitoring, pulse oximetry, end-tidal capnography, temperature, and airway pressure. Additional invasive monitoring, including direct arterial cannulation, central venous pressure, transesophageal echocardiography, and urinary catheterization may be warranted if the patient's cardiovascular status or the planned surgery may result in hemodynamic instability.11,20

One important caveat in the care of the patient with cyanotic heart disease is to be aware that conventional pulse oximetry overestimates arterial oxygen saturation as saturation decreases, and the discrepancy worsens with severe hypoxemia.21,25 Also, in the presence of right-to-left shunts, the end-tidal carbon dioxide readings consistently underestimate PaCO2, and this discrepancy also worsens with hypoxemia. The lower end-tidal carbon dioxide is explained by the relatively large dead-space ventilation caused by the right-to-left shunt.20,27,28 Arterial blood gases were obtained before induction and after induction and abdominal insufflation to assess differences in arterial and monitor values. This allowed a more accurate interpretation of monitor data.

Summary

Anesthesia providers must be prepared to handle hemodynamic and pulmonary instability quickly and effectively for the patient with cyanotic heart disease. Due to the improving morbidity and mortality of these patients, anesthetists are likely to be presented with an un repaired or repaired defect for noncardiac surgery. Careful planning and application of physiologic and pharmacologic principles will provide for optimum anesthetic management.

REFERENCES


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