Anesthesia for the Adult Patient With an Unrepaired Congenital Cyanotic Heart Defect: A Case Report

Marianne S. Cosgrove, CRNA, DNAP, APRN

Adult congenital heart disease, previously considered a rare comorbidity, is increasingly becoming a reality for today’s anesthesia providers. Improvements in prenatal diagnosis, sophisticated surgical techniques and equipment, advances in pediatric critical care, enhanced efficacy of cardiovascular pharmacologic agents, and an overall increase in postrepair survival rates have resulted in an estimated population of approximately 800,000 adults with congenital heart disease. Despite successful surgical repair or palliation, these individuals present the anesthesia provider with a multitude of challenges. Individualized care of these fragile patients should be approached with a keen understanding of the patient’s underlying cardiac anomaly.

This case report chronicles the anesthetic care of a 36-year-old woman presenting for left-sided ureteroscopy with laser lithotripsy and stent placement. Her medical history was remarkable for the presence of complex congenital heart disease consisting of multiple anomalies: a double-outlet right ventricle, transposition of the great arteries, pulmonary stenosis, atrial septal defect, and a hypoplastic left ventricle with concomitant mitral valve atresia.

General anesthesia was successfully administered, with meticulous attention given to maintenance of systemic vascular resistance to minimize shunting, oxygenation, administration of preprocedure antibiotics, and judicious replacement of intravenous fluids via air-filtered tubing.

Keywords: Adult, congenital heart disease, cyanotic heart lesions, general anesthesia.

It is difficult to accurately estimate the actual incidence rate of congenital heart disease. This is secondary to widespread variations in the scope and severity of its various forms. Congenital heart defects may present as mild lesions such as minor endocardial cushion defects or moderate to severe defects arising from morphologic anomalies of the myocardium and cardiac vasculature. In 2002, a meta-analysis of 44 studies attempted to ascertain a valid rate of congenital heart disease currently. As a result of this compilation of data, the incidence rate of moderate to severe congenital heart disease was estimated to range from 6 to 19 per 1,000 live births, with an overall rate of 75 per 1,000 live births. Congenital heart defects account for 50% of all infant deaths and approximately 33% of all hospitalizations secondary to inborn anomalies in the United States.

Although congenital heart disease was previously thought to be confined to the pediatric population, the prevalence of adult congenital heart disease (ACHD) continues to escalate. Interventions such as improved prenatal detection and diagnosis, advanced surgical techniques, novel medical therapies, and more efficacious pharmaceutical agents have facilitated the expansion of this subset of patients. Estimates of the prevalence of ACHD range from 500,000 to 800,000 to 1.5 million US adults. The growth of this population is expected to continue at a rate of 5% per year. It is now projected that 80% to 85% of patients with congenital heart disease will survive to adulthood. Due to the unique needs of these patients, there is currently a movement for a focused specialization on care of adult patients with congenital heart disease, as well as the development of regional specialty centers. A multidisciplinary team approach involving specialists in internal medicine, cardiology, cardiac surgery, nursing, and social services is best suited to the comprehensive care of the adult patient with congenital heart disease. These congenital lesions carry not only multisystemic manifestations but also psychosocial ramifications. With the advent of advanced medical and surgical techniques, the possibility of caring for the adult patient with unrepaired or palliated congenital heart disease, albeit rare, is present nonetheless. Due to the presence of a relatively small patient population coupled with complex medical demands and precarious conditions, it is alleged that adult patients with congenital heart disease may be prone to receive “suboptimal medical management.”

To date, there is a paucity of data regarding the anesthetic care of adult patients with unrepaired cardiac anomalies. Few of these patients survive into adulthood because of their serious physiological derangements. An expansive search of the literature provides only a few reports of patients with unrepaired cardiac lesions undergoing anesthesia for noncardiac surgery. Because
the effects of anesthesia on these special patients has not been extensively researched, specific, evidence-based recommendations for their anesthetic care continue to be developed.14

Case Summary
A 36-year-old, 62-kg woman presented emergently to the operating room with a symptomatic, 1-cm, distal left-sided ureterolithiasis. Her medical history was remarkable for complex congenital heart disease consisting of transposition of the great arteries, double-outlet right ventricle (RV), pulmonary stenosis without substantial pulmonary hypertension, atrial septal defect (ASD), and hypoplastic left heart syndrome (HLHS) with mitral atresia. She had been palliated with a balloon atrial septostomy at age 8 years, but the major constituents of her anomalies remained unrepaired. Additional surgical history included a cleft lip and palate repair at age 1 year and a cardiac ablation for chronic atrial flutter at age 28 years. She reported no untoward reactions to general anesthesia. Five years before her present admission, a symptomatic ureterolithiasis had been managed conservatively with intravenous (IV) hydration and analgesics. However, fulminant congestive heart failure quickly developed because of excessive fluid administration. Because of this history and the fact that the patient was experiencing severe discomfort, immediate surgical intervention with ureteroscopy, laser lithotripsy, and stent placement in the left ureter was indicated.

Preoperatively, the patient’s cardiologist was contacted by phone, and she verified that she had found the patient to be in stable condition 1 week before this hospitalization. After consultation between the urologic surgeon, the anesthesiologist, the Certified Registered Nurse Anesthetist (CRNA), the patient, and her father, it was decided to proceed with surgery under general endotracheal anesthesia. Spinal anesthesia was not considered as an option because of the potential for a precipitous drop in systemic vascular resistance (SVR), leading to an increase in right-to-left shunting, as well as because of the patient’s apprehension regarding the procedure.

The patient’s medical history was also notable for hypothyroidism and ventricular fibrillation and cardiac arrest at age 29 years, with sequelae of residual, mild neurologic deficit and long-term oxygen dependence. As a result, she received continuous oxygen therapy via nasal cannula at 4 L/min. She had an allergy to sulfa medications and a self-reported untoward reaction to epinephrine with a chief complaint of “rapid heart rate.” Her medication profile included amiodarone, 200 mg twice a day; digoxin, 0.125 mg 5 times a week; enalapril, 5 mg every day; ferrous sulfate, 325 mg every day; levothyroxine, 12.5 µg/d; and acetylsalicylic acid, 81 mg/d. Preoperative laboratory values were notable for a total white blood cell count of 18.1 × 1,000/µL (4.0-10.0 × 1,000/µL), differential neutrophils of 83% (38%-81%), differential lymphocytes of 7% (14%-46%), hematocrit of 45.2%, mean corpuscular volume of 96 fl (78-94 fl), creatinine of 1.3 mg/dL (0.6-1.2 mg/dL), aspartate aminotransferase of 43 U/L (0-35 U/L), bilirubin of 0.21 mg/dL (< 0.2 mg/dL), and 2+ proteinuria. Electrolytes, glucose, serum urea nitrogen, platelets, and international normalized ratio values were within normal limits.

The preoperative electrocardiogram revealed a left anterior hemiblock, right bundle branch block, left atrial enlargement, and left axis deviation. Auscultation of the precordium was remarkable for a loud holosystolic murmur. Chest radiograph was notable for cardiomegaly with enlarged pulmonary arteries centrally but with no active pulmonary disease. Her baseline vital signs were temperature, 36.4°C; pulse, 61 to 70/min; BP, 114 to 137/69 to 71 mm Hg; and arterial oxygen saturation (SaO₂), 88% to 89% on 4 L of oxygen administered via nasal cannula. Her airway was remarkable for the presence of a palatal obturator. She was classified with a Mallampati II score with good oral aperture, thyromental distance, and cervical range of motion. She reported a pain scale of 7 of 10 and had received morphine sulfate, 2 mg IV × 2, in the emergency department before being transferred to the operating room.

The patient was brought into the cystoscopy suite via stretcher. A 20-gauge IV catheter was inserted in her right wrist, and an infusion of lactated Ringer’s solution was initiated via a microdrip set. The IV tubing and stopcocks had been meticulously examined for the presence of air bubbles and subsequently de-aired. An inline air filter was added to the tubing, and all bolus IV medications were administered through it. Antibiotic prophylaxis was achieved with the slow IV administration of ampicillin, 2 g, and gentamicin, 80 mg, before commencement of the anesthetic induction and airway manipulation. Standard monitors were applied, and denitrogenation was achieved to an end-tidal nitrogen concentration of < 5% via sealed face mask. Oxygen saturation increased from baseline values to 98%. An end-tidal carbon dioxide (EtCO₂) concentration was obtained before anesthetic induction. It was noted to be lower than expected at 25 mm Hg due to the presence of the cyanotic lesion. Initial IV sedation consisted of midazolam, 1 mg, and fentanyl, 40 µg. Induction of anesthesia and muscle relaxation was achieved with ketamine, 60 mg IV, and rocuronium, 35 mg IV; the patient was mask ventilated before laryngoscopy, and the airway was found to be patent. She was intubated with ease on the first attempt with a 6.0-mm internal-diameter endotracheal tube. The smaller endotracheal tube was chosen because of the presence of a visibly small glottic aperture. Breath sounds were equal and clear to auscultation bilaterally. A modest increase in HR from 70/min to 81/min was noted with laryngoscopy and intubation; this resolved over the next 10 minutes.
without intervention and continued at baseline values for the remainder of the procedure.

Immediately after anesthetic induction, an arterial line was placed in the patient’s right radial artery to facilitate continuous monitoring of blood pressure (BP). The patient was placed in the lithotomy position. Anesthesia was maintained with 1% to 3% of sevoflurane and an inspired oxygen concentration (FIO2) of 0.97 to 1.0. The volatile agent was titrated according to BP and bispectral index values. A second dose of fentanyl, 20 μg IV, was administered during the course of the anesthetic, as well as 4 mg IV of ondansetron, as prophylaxis against the development of postoperative nausea and vomiting.

Vital signs remained stable throughout the course of the case, with BP ranging from 105 to 145/42 to 60 mm Hg; HR, 52/min to 81/min; SaO2, 94% to 98%; ETCO2, 31 to 40 mm Hg, and bispectral index, 51 to 58. The surgery proceeded without incident and was concluded within 45 minutes. Removal of the ureteral calculi was achieved with the use of a basket and was followed with left ureteral stent placement. Estimated blood loss was minimal, and total IV fluids were 400 mL: 200 mL of maintenance fluids and 200 mL for antibiotic administration. Residual and total IV fluids were 400 mL: 200 mL of maintenance fluids and 200 mL for antibiotic administration. Residual neuromuscular blockade was reversed with the slow administration of neostigmine, 2 mg, and glycopyrrolate, 0.4 mg IV. No apparent change in heart rate was appreciated subsequent to the administration of these agents. Emergence from anesthesia was smooth, and the patient was extubated awake after responding appropriately to commands.

The patient was transported to the postanesthesia care unit (PACU) and given 4 L/min of oxygen via nasal cannula in stable condition. Initial vital signs in the care unit (PACU) and given 4 L/min of oxygen via nasal cannula in stable condition. Estimated blood loss was minimal, and total IV fluids were 400 mL: 200 mL of maintenance fluids and 200 mL for antibiotic administration. Residual neuromuscular blockade was reversed with the slow administration of neostigmine, 2 mg, and glycopyrrolate, 0.4 mg IV. No apparent change in heart rate was appreciated subsequent to the administration of these agents. Emergence from anesthesia was smooth, and the patient was extubated awake after responding appropriately to commands.

After consultation with the surgeon, the anesthesiologist, the patient, and her father, it was concluded that she was in stable condition, and she was cleared for discharge. This decision was motivated both by the surgeon’s wish to circumvent the possibility of the patient contracting a nosocomial infection as well as the patient’s and her family’s strong desire to return home as quickly as was feasible. The patient was discharged to home in stable condition approximately 3 hours after her admission to the PACU. A postoperative follow-up call was made 24 hours after discharge by a same-day surgery PACU nurse, who found the patient to be free of complaints and reportedly feeling “back to normal.” No apparent surgical or anesthetic sequelae were noted.

Discussion

A review of the literature yields several case reports of adults with unrepaired congenital heart disease undergoing anesthesia for noncardiac surgery. One account describes the anesthetic care of a 24-year-old female patient with unrepaired tetralogy of Fallot undergoing laparoscopic cholecystectomy for pancreatitis. Although previously palliated with a modified Blalock-Taussig (right subclavian to right pulmonary artery) shunt, her tetralogy of Fallot remained unrepaired in terms of the essential components. A different case study reviews the successful anesthetic management of a 22-year-old woman undergoing appendectomy. She suffered from situs solitus, a severe cyanotic heart defect consisting of pulmonary atresia, overriding aorta, concordant atrioventricular shunts, aortopulmonary collateral circulation, and a large ventricular septal defect (VSD). Both a 22-year old man who received anesthesia for a repeated craniotomy to repair a chronic brain abscess and a 50-year-old woman anesthetized for emergency laparotomy presented with complex cyanotic congenital heart disease, further complicated by the presence of Eisenmenger syndrome. Eisenmenger syndrome, a severe pulmonary vascular disease, is the result of the longstanding communication between the pulmonary and systemic circulations. Chronic pulmonary vascular congestion occurs from substantial left-to-right shunting through an ASD or VSD. This results in the eventual production of pulmonary hypertension. As pulmonary hypertension increases, shunting ultimately reverses to a right-to-left direction, thereby causing cyanosis.

Common denominators in all aforementioned accounts were anesthetic plans that highlighted similar interventions: maintenance of intravascular volume and preload and avoidance of precursors to acidosis such as hypothermia, hypercarbia, and hypotension. Minimization of intracardiac shunting was typically achieved via avoidance of decreases in systemic vascular resistance and increases in pulmonary vascular resistance (PVR). All reported anesthetics were delivered successfully without evidence of anesthetic sequelae.

In an effort to comprehend the magnitude and physiologic implications of this patient’s structural cardiac defects, it is essential to understand and compare them with normal cardiac anatomy (Figure, left).

The first of the major anomalies present was transposition of the great arteries. Simple transposition is the most common cyanotic abnormality in the newborn. Major findings associated with this defect include an aorta emanating directly from the RV and a pulmonary artery that arises from the left ventricle. Because of the complete separation of the pulmonary and systemic circulations, mixing of the 2 is wholly dependent on the presence of 1 or more communicating intracardiac shunts, such as patent ductus arteriosus or ASD. Atrial
septal defects may be enlarged via a balloon atrial (also known as Rashkind) septostomy. This procedure, which the patient had undergone as an 8-year-old, is most frequently performed with the goal of facilitating systemic oxygenation in patients with cyanotic congenital heart disease.24

It is interesting to note that transposition of the great arteries may frequently accompany double-outlet RV,25 an additional lesion affecting this patient. Double-outlet RV is a defect in which the great vessels arise either predominantly or entirely from the RV.4,25,26 The RV may or may not be septated. Tricuspid regurgitation may also be present.26 Additionally, double-outlet RV is always associated with the presence of a VSD27; however, this particular feature was absent in this patient primarily due to the presence of HLHS.

Concomitant HLHS with mitral atresia contributed appreciably to this patient’s complex heart defect. With an incidence rate of 1.5 to 2 per 10,000 live births, HLHS represents the most common left-sided obstructive heart disease presenting in newborns. The heralding feature is mitral valve stenosis or complete atresia in concert with a severely underdeveloped left ventricle.28

In this patient, the combination of these anomalies essentially resulted in the presence of a 3-chambered heart, constituting a cyanotic lesion (Figure, right). Surgical correction of the transposition of the great arteries was not compulsory secondary to the existence of a single ventricle because mixing of pulmonary and systemic circulations was facilitated by the atrial septostomy. It was postulated that this patient’s systemic perfusion pressures and oxygen saturation remained relatively stable because of the preferential shunting of oxygenated blood past the absent mitral valve, across the large ASD, into the right atrium through the tricuspid valve, into the dilated RV, through the aortic valve, and into the aorta. Perfusion was also present through the stenotic pulmonary valve, presumably to lesser degree than the aortic valve.

Collectively with the mixing of oxygenated and deoxygenated blood, pulmonary stenosis shared a role in the production of cyanosis that was clearly evident in this patient. This stenosis may actually have played a “protective” role in that, despite the fact that pulmonary hypertension and Eisenmenger syndrome are frequently associated with ACHD as a result of chronic pulmonary vascular congestion,21 it was largely absent in this patient. It was the opinion of her cardiologist that this particular factor was probably most instrumental in her continued ability to function despite her array of major structural cardiac defects.

The RV has a tremendous capacity to adapt to volume and pressure demands. Although functionally of less importance than the left ventricle, in rare instances such as this, the RV may be responsible for both pulmonic...
and systemic perfusion. Symptoms of cardiac failure develop after RV dysfunction is present; this, in turn, is associated with major decreases in cardiac output and considerable morbidity and mortality. Moreover, due to the anatomical changes present with right-ventricular hypertrophy magnified by double-outlet RV syndrome, physical derangement or malposition of the atrioventricular node and His-Purkinje fibers are often noted. This morphologic alteration of the right side of the heart is a causative factor in these patients’ propensity to arrhythmias, most frequently supraventricular arrhythmias such as atrial flutter and fibrillation. Supraventricular arrhythmias are noted to exist more frequently in the following populations: patients with complex congenital heart disease, those whose functional defects have not been repaired, and those with chronic cyanosis. Additionally, the incidence of supraventricular arrhythmias is directly proportional to the age of the adult patient with congenital heart disease. Bundle branch blocks and lethal ventricular arrhythmias are common in patients with right-sided heart disease. The presence of a single ventricle is acknowledged to predispose the patient with congenital heart disease to a significantly greater incidence of cardiac standstill and sudden death. The patient in this case report had suffered a cardiac arrest at the age of 29 years, but no ectopy had been noted during her intraoperative or postoperative course.

Chronic cyanosis produces a specific array of physiologic effects, many of which were noted in this patient. Derangements of the hematologic system may include polycythemia, erythrocytosis, and attendant hyperviscosity as a result of the increased production of erythropoietin from chronic tissue hypoxia. Thromboembolic events are common, and IV hydration to decrease viscosity must be carefully administered to circumvent fluid overload and eventual congestive heart failure. Patients are prone to both procoagulation and anticoagulation abnormalities. These may develop in the presence of hepatomegaly and splenomegaly resulting from chronic vascular congestion. Thrombocytopenia and decreases in von Willebrand factor, factor V, and vitamin K-dependent factors may all be present in the cyanotic adult patient with congenital heart disease. Portal hypertension from hepatomegaly may induce a neurohormonal activation; this causes a retention of intravascular volume and the development of third spacing and exacerbation of edema. Hyperuricemia is frequently present and may develop as a result of uric acid overproduction and reduced clearance. Hyperuricemia predisposes the cyanotic patient to the development of nephrolithiasis, ureterolithiasis (as was noted in this patient), and gouty arthritis. The patient also exhibited proteinuria, which may be noted as a result of increased renal vessel pressure and congestion. Vessel proliferation throughout the body is the result of chronic cyanosis and increases in arteriolar wall stress. Attendant with hyperviscosity, both of these attributes stimulate the release of prostaglandins and nitric oxide, causing arteriolar dilation and eventual vascular congestion. Overall vessel engorgement is further mitigated by high central venous pressures. The summative effect of these factors place the cyanotic patient at high risk for the development of intraoperative bleeding. Primary side effects of this bleeding diathesis may range from mild symptoms such as hemoptysis and oozing from line insertion sites, to severe complications such as pulmonary or cerebral hemorrhage or both.

To date, there is no conclusive anesthetic plan that may be offered in the care of these patients. Preoperatively, an understanding of the patient’s underlying pathophysiology, coupled with knowledge of the effects of anesthesia on the anomalous myocardium, is crucial. Although extensive preoperative evaluations by the patient’s internist, cardiologist, and cardiac surgeon are preferable, they may not be readily available, as was the case with this patient. Careful review of the patient’s preoperative medication profile, with attention to potential anesthetic interactions, must be considered.

Bacterial endocarditis has posed a major concern for the patient with congenital heart disease; however, antibiotic prophylaxis has not been proved to prevent this occurrence and is currently recommended for use only in unrepaired or cyanotic forms of congenital heart disease. Because of the tendency of the patient with ACHD toward endocarditis, sterile technique during all invasive procedures should be ensured. Airway anomalies have been associated with certain forms of congenital heart disease; intubation of these patients may prove to be difficult, and preparation for this is encouraged. An inhalation induction with sevoflurane may be suitable for the adult with congenital heart disease, as it produces little direct myocardial depression. Additionally, sevoflurane is devoid of unwanted sympathomimetic effects or airway irritation, as is noted with the use of higher concentrations of desflurane. However, inhalation induction of anesthesia may be prolonged in the presence of pulmonary stenosis or right-to-left shunting. Conversely, IV induction of anesthesia may be prolonged in the presence of left-to-right shunting.

Ketamine has gained popularity for the IV induction of anesthesia because of its preservation of SVR and its bronchodilatory effect, and it was chosen for the IV induction of anesthesia in this patient. The use of ketamine for the induction of anesthesia “is considered one of the safest induction techniques for the cyanotic patient.” The combined effects of this agent are presumed to decrease the occurrence of right-to-left shunting during the induction phase of anesthesia. However, ketamine has been shown to exhibit direct myocardial depressant effects in the patient with a compromised myocardium. Etomidate, well known for its maintenance of cardiovas-
cicular stability, may be used for the induction of anesthesia, yet transient adrenocortical suppression following its use may contribute to the patient’s instability well into the postoperative period.\textsuperscript{37,38} Opioids such as fentanyl and sufentanil offer substantial cardiac stability and blunt the neuroendocrine response to surgical stress.\textsuperscript{37,38} Nevertheless, their overuse may lead to hypoventilation and concomitant untoward effects due to hypoxemia and hypercarbia,\textsuperscript{14,34} particularly in the postoperative period. This, in turn, may ultimately lead to an increase in PVR and the onset of right-to-left shunting. Muscle relaxants such as vecuronium and rocuronium have high autonomic margins of safety,\textsuperscript{37,38} but their reversal with anticholinesterase-anticholinergic combinations should be undertaken with care in an effort to avoid both arrhythmias and bradycardia.\textsuperscript{38}

With regard to the volatile anesthetic agents, it is known that they have little overall effect on PVR.\textsuperscript{37} However, the use of nitrous oxide (N\textsubscript{2}O) has been shown to increase PVR.\textsuperscript{15,34} Therefore, this agent was avoided in this patient. Additionally, due to its soluble nature, N\textsubscript{2}O may rapidly expand air bubbles, which may be inadvertently infused IV or entrained via the surgical site. These emboli represent a major concern in the patient with congenital heart disease, especially for those with right-to-left shunts, as the occurrence of a cerebral embolic event may have devastating consequences.\textsuperscript{39} Care must be taken with the preparation of all IV tubing and during the injection of IV medications; air filters should be used for both. Transesophageal echocardiography may be used intraoperatively to assess overall cardiac function, fluid status, and the presence of air emboli.\textsuperscript{17} Vasodilators as well as high concentrations of volatile anesthetics may exacerbate cyanosis by drastically decreasing SVR, thereby accentuating intracardiac shunting,\textsuperscript{13} particularly in single-ventricle patients.\textsuperscript{13} Phenylephrine may be used to reverse intraoperative hypotension. Increases in SVR from direct \(\alpha\)-stimulation and vasoconstriction will diminish the occurrence of right-to-left shunting.\textsuperscript{36} Cardiotoxic agents such as epinephrine and isoproterenol along with anticholinergics such as atropine may be employed in instances of attenuated cardiac output or standstill. However, the avoidance of tachydysrhythmias resulting from the use of these agents is of great importance. Maintenance of normothermia is also critical, as hypothermia and shivering greatly increase oxygen consumption and the demand on the myocardium. Sustained hypothermia may lead to decreased tissue perfusion via vasoconstriction, acidosis, and an increased PVR, which favors the development of right-to-left shunting.\textsuperscript{14}

Intraoperatively, vigilant monitoring of the patient with cyanotic heart lesions is essential. Continuous measurement of arterial BP may be achieved through the insertion of an arterial catheter. The assessment of central venous pressure may also be useful in monitoring intra-vascular fluid status. Concentrations of $\text{ETCO}_2$ may be inaccurate; lower-than-actual values may be noted in the presence of increased right-to-left shunting. Decreases in $\text{SaO}_2$ may signify increased right-to-left shunting produced by increasing PVR.\textsuperscript{14,34} Increases in PVR ultimately cause an aggravation of cyanosis and should be avoided at all costs. Increased PVR may occur from myriad factors, specifically hypoxia, hypercapnia, elevated airway pressures, respiratory and metabolic acidosis, and hypothermia.\textsuperscript{36,38} Overzealous manipulation of the airway may lead to bronchospasm; this may also contribute toward the development of increased PVR. Pulmonary hypertensive crisis resulting from increases in transthoracic pressures may lead to acute right heart failure and result in a major decrease in cardiac output,\textsuperscript{14,34} particularly in patients with single-ventricle disease. The susceptibility of patients with ACHD toward the development of pulmonary hypertension and congestive heart failure from considerable increases in PVR poses a real concern for the anesthesia provider. Pulmonary hypertensive crisis constitutes a major risk for adult patients with congenital heart disease, predisposing them to a significantly increased incidence of perioperative morbidity and mortality.\textsuperscript{14,34} Suggested modalities for the treatment of this occurrence are hyperventilation with an inspired oxygen concentration (Fi\textsubscript{O\textsubscript{2}}) of 1.0, judicious administration of volume, use of vasoconstrictors to elevate SVR, and control of wide variations of the heart rate.\textsuperscript{14,34,36}

**Conclusion**

Adult patients with congenital heart disease present the anesthetist with unique and varied challenges. Individualized care of these fragile patients should be approached with a keen understanding of the patient’s underlying cardiac anomaly, a focus on the preservation of oxygenation, and minimization of intracardiac shunting through careful pharmacologic manipulation of the circulation. Cautious administration of sedatives and anesthetic agents, control of wide variations in heart rate, avoidance of volume overload and subsequent congestive heart failure, and avoidance of IV air embolization is also recommended. Although previously rarely encountered in daily practice, ACHD will likely become more prevalent in the surgical population. With careful planning by the surgical and anesthesia care team, adult patients with congenital heart disease may be successfully anesthetized for noncardiac surgery without incident.

**REFERENCES**

3. Connor JA, Jenkins KJ. Factors associated with increased resource utilization for congenital heart disease. In: Wyszynski DF, Correa-