

CASE REPORT

Perioperative management of an adolescent with transposition of the great arteries, status post Senning procedure, for posterior spinal fusion

Nicole Elsey, MD¹, Joseph D. Tobias, MD^{1,2}

1. Departments of Anesthesiology, The Ohio State University, Columbus, Ohio (USA)

2. Departments of Anesthesiology & Pediatrics², Nationwide Children's Hospital, Columbus, Ohio (USA)

Correspondence: Joseph D. Tobias, MD, Chairman, Department of Anesthesiology & Pain Medicine, Nationwide Children's Hospital, 700 Children's Drive, Columbus, Ohio 43205 (USA); Phone: (614) 722-4200; FAX: (614) 722-4203; E-mail: Joseph.Tobias@Nationwidechildrens.org

ABSTRACT

With advances in surgical techniques and perioperative care, the survival rate of patients with congenital heart disease continues to increase. As such, patients with palliated or corrected congenital heart disease may present for major surgical procedures. Although alternative techniques are now available for the anatomic repair of patients with transposition of the great arteries (TGA), an atrial level baffle (Mustard or Senning procedure) was previously performed. As these patients age, long-term adverse effects may occur including myocardial dysfunction, rhythm disturbances requiring pacemaker placement, pulmonary hypertension, and baffle obstruction. We present a case report of a 14 year old girl with TGA, who underwent a Senning procedure during infancy and now presented for posterior spinal fusion in the treatment of kyphoscoliosis. The perioperative care of such patients is discussed.

Key Words: Congenital heart disease; Transposition of the great arteries; Senning procedure; Mustard procedure; Myocardial dysfunction; Pulmonary hypertension; Baffle obstruction

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INTRODUCTION

Congenital heart disease has a reported incidence of approximately 0.8 per 100 births.^{1,2} As such, patients with palliated or corrected congenital heart disease (CHD) may present for major surgical procedures. Although the goal of current surgical approaches is to achieve an anatomic repair, prior to the current advances in techniques, surgical procedures frequently resulted only in palliation. In such patients, appropriate attention to detail during the preoperative work-up, as well as the intraoperative and postoperative care can facilitate a successful perioperative course. We present a patient with transposition of the great arteries (TGA) who had undergone a Senning procedure during infancy who now presents for posterior spinal fusion (PSF) in the treatment of kyphoscoliosis. The perioperative care of such patients is discussed.

CASE REPORT

A 14 year old girl with TGA, who underwent a Senning procedure during infancy had now presented for posterior spinal fusion in the treatment of kyphoscoliosis. Review of her medical record and presentation of the material in this format were approved by the Institutional Review Board of Nationwide Children's Hospital, Columbus, Ohio. Her past history was significant for D-transposition of the great vessels diagnosed shortly after birth. During infancy, she had a Senning procedure. At 3 years of age, a VVI pacemaker was placed due to intermittent 3rd degree heart block. Although she had no other major medical problems, her mother stated that she was taking frequent afternoon naps due to fatigue and did not play competitive basketball this year as she could not keep up with her friends. Current medications included

sotalol 40 mg twice a day and enalapril 5 mg twice a day. Preoperative evaluation revealed a well developed, well nourished adolescent in no acute distress. Vital signs revealed an irregular rhythm with a heart rate of 84 beats/minute, blood pressure (BP) of 104/68 mmHg, respiratory rate of 16 breaths/minute, and a room air oxygen saturation of 97%. Physical examination was unremarkable. Echocardiography revealed no evidence of baffle leak or occlusion, the right ventricular cavity was slightly dilated with minimal compression of the left ventricular outflow tract. There was moderate tricuspid regurgitation and trace mitral regurgitation. There was moderate global depression of right ventricular function. Preoperative electrocardiograph revealed the presence of pacemaker spikes. Recent Holter monitoring revealed that the patient was pacemaker dependent less than 5% of the time. Preoperative laboratory evaluation revealed a hemoglobin value of 14.2 gm/dL, a hematocrit of 40.8%, and a platelet count of 312,000/mm³. The prothrombin time was 13.8 seconds, the INR was 1.1, and the partial thromboplastin time was 33 seconds.

A discussion with the pediatric cardiology service was held regarding appropriate pacemaker care during the surgical procedure. They were concerned that the use of a magnet and conversion of the pacemaker into a VOO mode would lead to the risk of arrhythmia induction related to an R on T phenomenon. Therefore, their recommendation was to leave the pacemaker in the VVI mode during the case. Since the patient was pacemaker dependent less than 5% of the time, they were not concerned regarding cautery interference. The patient was kept *nil per os* for 6 hours. Her morning doses of sotalol and enalapril were held. She transported to the operating room where routine monitors per the American Society of Anesthesiologists guidelines were placed. Given the patient's anxiety, inhalational induction was carried out with gradual increases in the inspired concentration of sevoflurane in 70% nitrous oxide in oxygen. When an appropriate depth of anesthesia was achieved, a 16G intravenous cannula was placed in the left forearm. Tracheal intubation was facilitated with sufentanil 50 µg and a single dose of rocuronium 50 mg. Antibiotic prophylaxis was done with cefazolin (2 gm) IV. Following tracheal intubation, a second peripheral intravenous cannula, a radial arterial cannula, and a double lumen central venous catheter were placed. The baseline CVP was 10-12 mmHg. Maintenance anesthesia consisted of desflurane (expired concentration 4%) and a sufentanil infusion adjusted to maintain the mean arterial pressure at 55-65 mmHg. No additional doses of neuromuscular blocking agents were administered during the case. Electrodes were placed for neurophysiologic monitoring (motor and somatosensory

evoked potentials). Test stimulation for motor evoked potentials (MEP's) revealed that adequate potentials could be obtained with 450 mV without interference with the pacemaker. The patient was turned prone onto a Jackson table for the procedure. The pacemaker module was noted with the patient turned prone and appropriate access ensured should it be necessary to place a magnet on it during the case. A magnet was kept ready on the anesthesia cart during the case. The sufentanil infusion rate was increased to a maximum of 0.7 µg/kg/hr to maintain the MAP at 55-65 mmHg. During use of the cautery for dissection of the paraspinous muscles, there was occasional interference with the pacemaker and loss of function; however, the patient's HR was ≥ 60 beats/minute throughout the case. The duration of the surgical procedure was 4.5 hours. Mixed venous oxygen saturation measured from the CVP cannula varied from 65-78% during the case. Intraoperative blood loss was approximately 500 ml. Fluid administration included a total of 2200 ml of lactated Ringers solution to maintain the CVP at 8-12 mmHg. Urine output during the case was 950 ml. A single dose of sodium bicarbonate (50 mEq) was administered for a base deficit of -6. The final hemoglobin was 10.9 gm/dl with a hematocrit of 32%. Sixty minutes before the completion of the surgical procedure, the sufentanil infusion was discontinued and a total of 1 mg of hydromorphone was administered in increments of 0.2 mg based on the patient's respiratory rate. Upon completion of the surgical procedure, the desflurane was discontinued, and the patient was turned supine. She spontaneously opened her eyes and moved all extremities in response to painful stimuli. Her trachea was extubated and she was transferred to the post-anesthesia care unit. Postoperative analgesia was provided by patient-controlled analgesia with hydromorphone. Supplemental oxygen at 2 lit/min was administered using nasal cannula for the first 24 postoperative hours. Her routine medications of sotalol and enalapril were restarted on first postoperative day. She was transitioned to oral analgesics (hydrocodone with acetaminophen) on third postoperative day and discharged home on 4th postoperative day.

DISCUSSION

TGA, classified as a cyanotic congenital cardiac malformation, exists when the aorta originates from the right ventricle and the pulmonary trunk arises from the left ventricle. TGA is divided into dextro-looped (D-TGA) and levo-looped (L-TGA) based on whether the atria and ventricles are concordant or discordant, respectively.³ Although L-TGA, also known as congenitally corrected transposition, is a

rare defect accounting for approximately 0.5% of all congenital cardiac malformations, D-TGA is the second most common congenital cardiac defect noted at birth affecting 1:3500-500 live births with a male:female ratio of 3.2:1.³ With D-TGA, a morphological right atrium is connected to a morphological right ventricle which gives rise to the aorta. A morphological left atrium is connected to a morphological left ventricle which gives rise to the pulmonary artery. This combination of concordant atrioventricular and discordant ventriculo-arterial connections creates two parallel circulation systems in which oxygenated blood recirculates within the pulmonary circuit via the left ventricle and pulmonary trunk while deoxygenated systemic blood recirculates to the body via the right ventricle and aorta.³ As compared to a physiologically normal circulation system that exists in series, the parallel circuits that are present with D-TGA result in systemic cyanosis.

Postnatal survival of an infant with D-TGA is dependent on the presence of intracardiac mixing of blood via an atrial septal defect (ASD), a patent foramen ovale (PFO), or a ventricular septal defect (VSD).³ Infants with intact atrial and ventricular septa are cyanotic at birth and rapidly decompensate due to inadequate mixing of the blood from the two parallel circulations. Palliative treatment with immediate balloon or surgical septostomy is necessary to increase mixing of blood at the atrial level.³ Prostaglandin E infusions may be used as an adjunctive therapy to maintain ductal patency, thereby increasing the left-to-right shunting of blood across the ductus arteriosus.³ The increased blood from the aorta across the ductus arteriosus to the pulmonary artery increases the flow of blood across the lungs into the left atrium. The increased left atrial return increases left atrial pressure, thereby facilitating the mixing of blood at the atrial level. Neonates with D-TGA and an associated ASD or VSD may be only mildly cyanotic at birth. Symptoms may be overlooked until 2-6 weeks of age, at which time changes in the pulmonary vascular resistance occur, resulting in the development of congestive heart failure.³ Initially, peripheral vascular resistance (PVR) is greater than, or equivalent to the systemic vascular resistance, allowing partial mixing of blood between the parallel systemic and pulmonary circuits via the ASD or VSD. As PVR falls, the balance is disturbed thereby allowing increased shunting from the systemic to the pulmonary circuit resulting in pulmonary congestion and congestive heart failure. At that time, the infants often present with tachypnea, tachycardia, and mild systemic cyanosis.³

The Senning operation for TGA was first reported by Senning in 1959, with later work by Quaegebeur in the late 1970's.^{4,5} It became the operation of choice for

surgical repair of TGA during this time period. The Senning procedure uses a 'baffle' between the atria resulting in the redirection of deoxygenated vena caval blood through the mitral valve to the morphological LV and pulmonary arteries and oxygenated pulmonary venous blood through the tricuspid valve into the morphological RV and aorta.^{6,7} The 'baffle' created in the Senning procedure is derived from autologous atrial tissue that is cut and folded in a complex manner to allow for the redirection of blood flow.⁷ Because of the technical difficulty associated with the formation of the Senning 'baffle', this procedure was not widely embraced. In 1964, Mustard simplified the procedure by excising the atrial septum and placing a 'single, synthetic pantaloons-shaped patch' between the atria to create the 'baffle', again redirecting the blood flow in the same manner as the Senning procedure.⁷ However, neither the Senning nor the Mustard procedures were without long term complications. Overall, the survival rate for patients following hospital discharge was 84-95% at 10 years and 76-89% at 15-20 years, with an actuarial decline in survival of approximately 0.5% per year.⁸

Sinus node dysfunction is one of the most common late complications after atrial-level repair with only 40% of patients remaining in sinus rhythm 15-20 years post-surgery.^{6,7} The suspected etiology of this complication is either damage to the sinus node artery during surgery or the progressive development of fibrosis at the atrial surgical scar lines.⁶ A junctional escape rhythm is common in patients with sinus node dysfunction thereby limiting the normal increase in heart rate that is seen during activity.⁶ As a result, patients are at increased risk for the development of atrial arrhythmias and sudden cardiac death. Nearly 50% of post-surgical deaths are sudden and likely related to the abrupt onset of polymorphic ventricular tachycardia/fibrillation(VT/VF)^{6,8,9}

The management of sinus node dysfunction and atrial arrhythmias can be challenging. Both pacemaker placement and radiofrequency ablation can be technically difficult as a result of altered anatomy and development of atrial scar tissue, while anti-arrhythmic medications can precipitate complete heart block.⁷ Implantation of a pacemaker is recommended for children with advanced second-degree heart block or complete atrioventricular block, patients with sick sinus syndrome who are symptomatic or have functional limitations, and those with tachy-bradycardia syndromes with recurrent events despite antiarrhythmic medications.^{10,11} Symptomatic patients and those with a documented history of atrial fibrillation/flutter should undergo electrophysiologic studies for inducible VT/VF with placement of an implantable cardioverter-defibrillator

(ICD) in those that have a positive EP study.⁹

Right ventricular dysfunction is another well-recognized, late complication associated with the Senning or Mustard atrial-repair, as the morphologic RV serves as the systemic ventricle. The etiology of RV dysfunction is not known; however there are several postulated theories. The triangular shape of the RV, in comparison to the bullet-shaped LV, may render the RV inadequate to sustain the systemic workload.⁶ The orientation of the myocardial fibers may also contribute to RV dysfunction. The RV endocardial and epicardial fibers are oriented in an oblique manner to the direction of blood flow resulting in a peristaltic motion with contraction; as compared to the LV, in which endocardial and epicardial fibers are perpendicularly oriented with a zone of circular myocytes in between resulting in a wringing motion with contraction.^{12,13} This differentiation in myocyte orientation and contractile motion normally allows the LV to function against high-resistance systems.¹³

Distention of the RV with progressive dysfunction and annular dilation may be one contributing factor towards development of RV dysfunction; however, tricuspid regurgitation may precipitate it as well.⁷ Under high-pressure from the systemic circulation, the septal wall of the RV bows toward the low-pressure LV thereby pulling the septal leaflet of the tricuspid valve.⁶ Thus, coaptation of the valve leaflets is lost and the valve becomes incompetent.

Pulmonary hypertension can be another late complication of atrial-level repair with an incidence reported at 7% in those patients surviving to adulthood.^{6,7} The presence of a ventricular septal defect and D-TGA repair after 1 year of age are risk-factors for the development of pulmonary hypertension.⁶ Signs and symptoms of pulmonary hypertension often do not appear until adulthood, a result of the LV's capacity to function against a high-resistance system.^{6,7} Given the significant perioperative risk imposed by pulmonary hypertension, its identification is key to successful perioperative management. The specific perioperative concerns related to pulmonary hypertension have been extensively reviewed elsewhere.^{14,15}

As noted above, patients may develop several late complications from the Senning and Mustard procedures. With current cardiology care, many of these patients are living into the 2nd and 3rd decade of life. As such, there are an increasing number of patients who may require anesthetic care during various types of surgical procedures. Given the complex nature of these patients and the potential for co-morbidities related to the primary defect or acquired during life, a thorough

preoperative evaluation is necessary to ensure the safe perioperative care of such patients. The 3 major issues related to the anatomy of the Mustard or Senning procedure which should be addressed preoperatively include: 1) an evaluation of myocardial function given that the systemic ventricle is the right ventricle, 2) an evaluation for the presence of pulmonary hypertension, and 3) an evaluation of the baffle for obstruction or leaks. Initial evaluation of these patients should begin with a complete cardiac history, specifically questioning on exercise capacity or intolerance, orthopnea or paroxysmal nocturnal dyspnea, the presence of palpitations, a documented history of arrhythmia, and prior syncopal events.⁶ On physical examination, the precordium will be hyperdynamic owing to the systemic workload of the right ventricle. A holosystolic murmur is indicative of systemic atrioventricular valve regurgitation or residual VSD, while the presence of a systolic ejection murmur may indicate subpulmonary stenosis.

Common post-atrial repair ECG findings include sinus bradycardia or a junctional rhythm, evidence of single or dual-chamber pacing, and signs of right ventricular hypertrophy. RVH can be identified on ECG by the presence of right-axis deviation, large R-waves in leads V₁ and AVR, and evidence of right atrial enlargement. ST-segment depression and T-wave inversion in the right precordial leads may be indicative of severe RVH.^{6,7} If available, prior Holter monitor recordings should be evaluated for paroxysmal atrial fibrillation/flutter, frequent supraventricular extrasystoles, junctional escape rhythm at rest, or episodes of non-sustained ventricular tachycardia.

In those patients with a pacemaker or ICD in place, additional perioperative management and evaluation is required in order to prevent device-related adverse events, e.g. damage to the device or leads, device failure, alterations in pacing behavior, inappropriate delivery of a defibrillatory shock, and inadvertent conversion to backup pacing modes.^{16,17} Electromagnetic interference (EMI) is the most commonly encountered problem intraoperatively and can occur as a result of electrocautery use, nerve stimulators, evoked potential monitors, fasciculations, shivering, large tidal volumes, external defibrillation, magnetic resonance imaging, radiofrequency ablation, extracorporeal shock wave lithotripsy, and electroconvulsive therapy.^{16,17} Current perioperative guidelines on cardiac rhythm management devices established by the American Society of Anesthesiologists (ASA) are outlined in reference 16. Generally, in pacemaker-dependent patients, the device will be reprogrammed to an asynchronous mode during the perioperative period, and the anti-tachyarrhythmia function of an ICD will be disabled.

In our patient, after a discussion with the pediatric cardiology service, it was decided to leave the pacemaker in the VVI mode during the case. Since the patient was pacemaker dependent less than 5% of the time, there was a limited concern that EMI interference would result in profound bradycardia or no cardiac output. The concern with the use of a magnet and conversion of the pacemaker into a VOO mode was the risk of arrhythmia induction related to an R on T phenomenon. However, we made sure that after positioning we had access to the pacemaker and a magnet in the room in the event that its use became necessary. The other aspect of our case that was unique was the use of neurophysiologic monitoring including both somatosensory and motor evoked potentials (see below). As these involve electrical stimulation, we ran several test runs prior to turning the patient prone to verify that we could elicit the needed responses at a low stimulus threshold without interfering with pacemaker function.

Perhaps the most important aspect of the preoperative evaluation in a TGA patient status-post atrial switch is assessment of the systemic ventricular function. Echocardiography, either by transthoracic or transesophageal route, plays an essential role in the assessment of right ventricular structure, function, and physiology. If the patient's body habitus is amenable, transthoracic echocardiography (TTE) should be employed to evaluate for myocardial perfusion defects, uncoordinated myocardial contraction, tricuspid valve regurgitation, and the presence of an atrial baffle leak, all of which can contribute to RV failure.¹⁸ Hoffman et al demonstrated that the use of subcostal TTE imaging planes, instead of standard apical planes, allowed for the simultaneous assessment of the RV inflow and outflow tracts, as well as right ventricular function.¹⁹ When comparing RV performance during cardiopulmonary exercise and dobutamine stress testing, Li et al found that exercise capacity correlated with systemic ventricle free wall excursion at rest and during dobutamine stress.²⁰ Depressed RV function and exercise capacity may be the result of inducible ischemia, RV hypertrophy with increased myocardial oxygen demand or an inability to increase stroke volume as a result of fixed preload created by the resistance of the intra-atrial baffle.^{20,21}

The use of cardiac MRI has significantly reduced the need for TEE or cardiac catheterization. Gadolinium-enhanced MRI can be used to assess systemic right ventricle function and evaluate areas of abnormal myocardium.²² The presence of abnormal myocardial regions on MRI correlates with markers of adverse outcomes including RV dysfunction, poor exercise tolerance, presence of arrhythmias, and clinical deterioration over time. B-type natriuretic peptide

(BNP) belongs to a family of peptide hormones that are involved in the homeostasis of intravascular volume status, vascular resistance, and myocardial function. BNP is produced in the cardiac myocytes, is released into the circulation as a result of increased ventricular wall stress, and causes natriuresis and vasodilation to counter the effects of the rennin-angiotensin-aldosterone system (RAAS).²³⁻²⁵ BNP levels may also serve as a biomarker of disease severity with prognostic applicability in CHD patients. A stepwise increase in BNP has been shown to correlate with increasing disease severity and decreasing systemic ventricular function, such that a BNP value > 78 pg/mL had a high predictive accuracy in terms of overall mortality.²⁶ BNP levels as an indicator for disease severity may also be applicable in the prediction of outcomes for patients with pulmonary hypertension. Bernus et al found that changes in BNP over time correlated inversely with cardiac index and positively with mean RAP, mean pulmonary artery pressure, PCWP, and pulmonary vascular resistance index.²⁷

In addition to the evaluation of RV function, it is also important to evaluate these patients for the presence of pulmonary hypertension (PH) preoperatively. The most common presenting symptoms of PH include dyspnea on exertion, fatigue, chest pain, syncope, palpitations and lower extremity edema.^{28,29} A right heart catheterization (RHC), which is the gold standard for evaluation of pulmonary hypertension, should be considered. Several studies have demonstrated that children with PH have a significant risk of perioperative cardiac complications including cardiac arrest, pulmonary hypertensive crisis, and death.^{30,31} The perioperative management of patients with PH can be challenging. Patients on chronic therapy for PH including intravenous prostacyclins (epoprostenol and treprostinil), phosphodiesterase inhibitors (sildenafil), endothelin antagonists (bosentan), and inhaled prostacyclin analogs (iloprost) should be continued on these medications throughout the perioperative period.^{30,31} Intraoperatively, hypercarbia, alveolar hypoxia, systemic hypoxemia, metabolic acidosis, and noxious stimuli, such as pain or airway instrumentation, can trigger a rapid rise in PVR, and even a pulmonary hypertensive crisis.³³ Ventilation strategies to minimize pulmonary hypertensive triggers should be employed and include the use of high oxygen concentrations, low tidal volumes (6 mL/kg predicted body weight), a slightly elevated respiratory rate to allow for mild hypocarbia, and optimal levels of positive end-expiratory pressure (5-10 cmH₂O) to reduce the degree of atelectasis and maintain functional residual capacity. The medications used to induce and maintain anesthesia intraoperatively can also influence PVR and the management of PH.

The volatile agents attenuate hypoxic pulmonary vasoconstriction thereby increasing ventilation-perfusion mismatching. Isoflurane and sevoflurane are associated with pulmonary vasodilation and are generally accepted as safe anesthetic components in patients with PH. Furthermore, the use of benzodiazepines, opioids, nitrous oxide, etomidate, neuromuscular blocking agents, and propofol are regarded as safe for use in patients with pulmonary hypertension, as they have little to no effect on PVR. Moderate hyperventilation with 100% oxygen, along with the initiation of inhaled nitric oxide (iNO), are the first line of treatments for intraoperative PH.³²⁻³⁵

Senning and Mustard repair patients with both pulmonary hypertension and RV dysfunction may benefit from the perioperative use of milrinone. In patients who have tricuspid regurgitation (TR), in addition to RV dysfunction, milrinone reduces SVR while maintaining a normal-high heart rate. Perioperatively, the impact of medications with negative inotropic properties including volatile anesthetic agents and β -adrenergic antagonists should be considered.^{36,37}

In the prone position, there is an increase in CVP, a decrease in left ventricular end-diastolic diameter, and no overall change in systolic function.³⁸ As such, in patients with poor RV function undergoing surgery in the prone position, in which volume management is crucial for hemodynamic stability, the use of intraoperative TEE and CVP monitoring, continuous mixed-venous oxygen saturation (SvO₂) may be used as an indicator of cardiac output and oxygen consumption.^{39,40} The use of continuous SvO₂ monitoring can also serve as a guide to fluid and inotropic management with the demonstration that SvO₂-guided therapy may reduce both hospital stay and postoperative complications.^{41,43}

Intraoperative hemodynamic management may also be significantly affected by the patient's routine perioperative medications, more specifically angiotensin-converting enzyme inhibitors (ACE-I's) and angiotensin-receptor blockers (ARB's). These medications antagonize the renin-angiotensin-aldosterone system (RAAS) resulting in direct sympathetic blockade, increased bioavailability of intrinsic vasodilators, inhibition of angiotensin II, and reduced secretion of aldosterone and antidiuretic hormone.⁴⁴ The clinical significance, particularly related to anesthetic management, is the inhibition of angiotensin II, which normally functions as a potent vasoconstrictor and stimulus for arginine-vasopressin release; two substances which oppose the hypotensive effects of anesthetic agents.⁴⁵ As a result, patients treated with ACE-I's and ARB's in the perioperative setting are at higher risk of developing clinically significant hypotension after anesthetic induction.⁴⁶⁻⁴⁸

Successful treatment of RAAS antagonist hypotension requires adequate intravascular volume replacement and potentially the use of vasopressin as the hypotension may be unresponsive to adrenergic agonists such as phenylephrine.⁴⁶⁻⁴⁸ Although there remains limited consensus on the continuation or withdrawal of RAAS antagonists perioperatively, several studies have demonstrated an increased risk of refractory hypotension when ACE-I's and ARB's are administered on the morning of surgery.^{49,50} As such, the authors of these studies, as well as several institutions, recommend that patients on chronic ACE-I and ARB therapy receive their last dose of medication on the day before surgery. Given these concerns, our patient's usual dose of enalapril was not administered the morning of surgery and vasopressin was readily available in the operating room in the event of hypotension.

Prior to the initiation of surgery, it is also important to determine if the patient should receive bacterial endocarditis (SBE) prophylaxis.⁵¹ National Surgical Infection Prevention Project (NSIPP) and Centers for Disease Control (CDC) recommend the administration of prophylactic antibiotics to reduce the risk of surgical site infection.⁵² The use of intraoperative neuromonitoring, including somatosensory-evoked potentials (SSEP's) and motor-evoked potentials (MEP's), can further impact the anesthetic management of patients undergoing PSF. Because the blood supply to the spinal cord comes from two separate supplies (the anterior and posterior spinal arteries), MEP monitoring is used to assess the anterior spinal cord for injury while SSEP monitoring is used to assess the dorsal spinal columns.^{53,54} In general, MEP's are more sensitive to anesthetic agents than SSEP's with longer pathways being the most sensitive. Therefore, the most difficult signals to attain are lower extremity MEP's.⁵⁵⁻⁵⁷ Our usual practice as demonstrated by this patient is to use a single dose of an intermediate acting neuromuscular blocking agent to facilitate tracheal intubation and then allow for spontaneous recovery to permit MEP monitoring. Our intraoperative anesthetic regimen included 0.5 MAC desflurane and a sufentanil infusion.

Lastly, methods to reduce intraoperative blood loss, and in turn, complications associated with large volume blood loss and allogeneic blood transfusions should be addressed. The transfusion of allogeneic blood products is not without risk; the potential for transmission of infectious diseases, immunosuppression, transfusion-related acute lung injury, transfusion reactions, graft-versus-host disease, and increased risk of nosocomial infections and pneumonia have all been recognized as adverse events.⁵⁸⁻⁶²

Several techniques exist to both minimize the amount of blood loss and limit the need for allogeneic transfusions, and they include autologous transfusion therapy; intraoperative blood salvage; pharmacologic manipulation of the coagulation cascade utilizing epsilon-aminocaproic acid, tranexamic acid, desmopressin (DDAVP), or recombinant factor VIIa; and controlled hypotension.^{58,63} Proper patient positioning, especially when prone, to minimize impedance of venous return due to pressure on the abdomen, and maintenance of normothermia can further help to reduce blood loss.⁶⁴ Given our patient's stable hemodynamic status preoperatively, we chose to use sufentanil in doses as high as 0.5 µg/kg/min to maintain a MAP at 55-65 mmHg.

In summary, we present the perioperative considerations in caring for a patient with TGA who had undergone a Senning procedure during PSF. Perioperative concerns include not only those related to the primary cardiac lesion, but also those related to the surgical procedure and intraoperative neurophysiologic monitoring. Cardiac and hemodynamic concerns of these patients include the potential for RV dysfunction, baffle leaks or obstruction, the presence of pulmonary hypertension, and sinus node dysfunction with arrhythmias. The latter may necessitate placement of a pacemaker or ICD device. Depending on the patient's cardiac status, the impact of anesthetic agents on myocardial function, sinus node function, and PVR should be considered. Additional concerns include those related to PSF including blood conservation techniques and the need for neurophysiologic monitoring.

REFERENCES

- Mathews TJ, Miniño AM, Osterman MJK, et al. Annual summary of vital statistics: 2008. *Pediatrics* 2011;127:146-57.
- van der Bom T, Zomer AC, Zwinderman AH, Meijboom FJ, Bouma BJ, Mulder BJ. The changing epidemiology of congenital heart disease. *Nat Rev Cardiol* 2011;8:50-60.
- Anderson R and Weinberg P. The clinical anatomy of transposition. *Cardiol Young* 2005;15(suppl 1):76-87.
- Senning A. Surgical correction of transposition of the great vessels. *Surgery* 1959;45:966-80.
- Quaegebeur JM, Rohmer J, Brom AG. Revival of the Senning operating for treatment of transposition of the great arteries. *Thorax* 1977;32:517-24.
- Love BA, Mehta D, Fuster VF. Evaluation and management of the adult patient with transposition of the great arteries following atrial-level (Senning or Mustard) repair. *Nat Clin Pract Cardiovasc Med* 2008;5:454-67.
- Warnes CA. Congenital heart disease for the adult cardiologist: Transposition of the great arteries. *Circulation* 2006;114:2699-709.
- Dos L, Teruel L, Ferreira IJ, et al. Late outcome of Senning and Mustard procedures for correction of transposition of the great arteries. *Heart* 2005;91:652-6.
- Kammeraad J, van Deurzen C, Sreeram N, et al. Predictors of sudden cardiac death after Mustard or Senning repair for transposition of the great arteries. *J Am Coll Cardiol* 2004;44:1095-1102.
- Drago F, Silvetti M, Grutter G, De Santis A. Long term management of atrial arrhythmias in young patients with sick sinus syndrome undergoing early operation to correct congenital heart disease. *Europace* 2006;8:488-94.
- Villain E. Indications for pacing in patients with congenital heart disease. *Pace* 2008;31:s17-20.
- Haddad F, Couture P, Tousignant C, Denault A. The right ventricle in cardiac surgery, a perioperative perspective: I. Anatomy, physiology, and assessment. *Anesth Analg* 2009;108:404-21.
- Nielsen E, Smerup M, Agger P, et al. Normal right ventricular three-dimensional architecture, as assessed with diffusion tensor magnetic resonance imaging, is preserved during experimentally induced right ventricular hypertrophy. *Anat Rec* 2009;292:640-51.
- Carmosino MJ, Friesen RH, Doran A, Ivy DD. Perioperative complications in children with pulmonary hypertension undergoing noncardiac surgery or cardiac catheterization. *Anesth Analg* 2007;104:521-7.
- Strumpher J, Jacobsohn E. Pulmonary hypertension and right ventricular dysfunction: physiology and perioperative management. *J Cardiothorac Vasc Anesth* 2011;25:687-704.
- Stone M, Apinis A. Current perioperative management of the patient with a cardiac rhythm management device. *Semin Cardiothorac Vasc Anesth* 2009;13:31-43.
- Navaratnam M, Dubin A. Pediatric pacemakers and ICD's: how to optimize perioperative care. *Pediatr Anaesth* 2011;21:512-21.
- Haddad F, Couture P, Tousignant C, Denault A. The right ventricle in cardiac surgery, a perioperative perspective: II. Pathophysiology,

- clinical importance, and management. *Anesth Analg* 2009;108:404-21.
19. Hoffman P, Szymanski P, Lubiszewska B, Rozanski J. Echocardiographic evaluation of the systemic ventricle after atrial switch procedure. The usefulness of subcostal imaging. *Cardiol J* 2008;15:156-61.
 20. Li W, Hornung T, Francis D, et al. Relation of biventricular function quantified by stress echocardiography to cardiopulmonary exercise capacity in adults with mustard (atrial switch) procedure for transposition of the great arteries. *Circulation* 2004;110:1380-6.
 21. Budts W, Scheurwegs C, Stevens A, et al. The future of adult patients after mustard or senning repair for transposition of the great arteries. *Int J Cardiol* 2006;113:209-14.
 22. Giardini A, Lovato L, Donati A, et al. Relation between right ventricular structural alterations and markers of adverse clinical outcome in adults with systemic right ventricle and either congenital complete (after Senning operation) or congenitally corrected transposition of the great arteries. *Am J Cardiol* 2006;98:1277-82.
 23. Tobias J. B-type natriuretic peptide: diagnostic and therapeutic application in infants and children. *J Intensive Care Med* 2011;26:183-95.
 24. Knirsch W, Hausermann E, Fasnacht M, et al. Plasma B-type natriuretic peptide levels in children with heart disease. *Acta Paediatrica* 2011;100:1213-6.
 25. Trojnarowska O, Gwizdala A, Katarzynski S, et al. The BNP concentrations and exercise capacity assessment with cardiopulmonary stress test in cyanotic adult patients with congenital heart disease. *Int J Cardiol* 2010;139:241-7.
 26. Giannakoulas G, Dimopoulos K, Bolger A, et al. Usefulness of natriuretic peptide levels to predict mortality in adults with congenital heart disease. *Am J Cardiol* 2010;105:869-73.
 27. Bernus A, Wagner B, Accurso F. Brain natriuretic peptide levels in managing pediatric patients with pulmonary arterial hypertension. *Chest* 2009;135:745-51.
 28. Fischer L, Van Aken H, Burkle H. Management of pulmonary hypertension: physiological and pharmacological considerations for anesthesiologists. *Anesth Analg* 2003;96:1603-16.
 29. Blaise G, Langleben D, Hubert B. Pulmonary arterial hypertension. *Anesthesiology* 2003;99:1415-32.
 30. Carmosino M, Friesen R, Doran A, Ivy D. Perioperative complications in children with pulmonary hypertension undergoing noncardiac surgery or cardiac catheterization. *Anesth Analg* 2007;104:521-7.
 31. Friesen R, Williams G. Anesthetic management of children with pulmonary arterial hypertension. *Ped Anesth*. 2008;18:208-16.
 32. Mossad E. Pro: intraoperative use of nitric oxide for treatment of pulmonary hypertension in patients with congenital heart disease is effective. *J Cardiothorac Vasc Anesth* 2001;15:259-62.
 33. Dietrich C, Tobias J. Intraoperative administration of nitric oxide. *J Int Care Med*. 2003;18:146-9.
 34. Haddad E, Lowson S, Johns R, Rich G. Use of inhaled nitric oxide perioperatively and in intensive care patients. *Anesthesiology* 2000;92:1821-5.
 35. Forrest P. Anaesthesia and right ventricular failure. *Anaesth Intensive Care* 2009;37:370-85.
 36. Priebe H. Effects of halothane on global and regional biventricular performances and on coronary hemodynamics before and during right coronary artery stenosis in the dog. *Anesthesiology* 1993;78:541-52.
 37. Palmisano B, Mehner R, Baker J, et al. Direct effects of halothane and isoflurane in infant rabbit hearts with right ventricular hypertrophy secondary to chronic hypoxemia. *Anesth Analg* 1995;80:1122-8.
 38. Soliman DE, Maslow AD, Bokesch PM, et al. Transoesophageal echocardiography during scoliosis repair: comparison with CVP monitoring. *Can J Anaesth* 1998;45:925-32.
 39. Vincent J. Does central venous oxygen saturation accurately reflect mixed venous oxygen saturation? Nothing is simple, unfortunately. *Intensive Care Med* 1992;18:386-7.
 40. Naruns E. Clinical implications of intraoperative continuous mixed venous oxygen saturation monitoring. *AANA J*. 1989;57:45-52.
 41. Della Rocca G, Pompei L. Goal-directed therapy in anesthesia: any clinical impact or just a fashion. *Minerva Anestesiologica* 2011;77:545-53.
 42. Viale J, Annat G, Ravat F, et al. Oxygen uptake and mixed venous oxygen consumption during aortic surgery and the first three postoperative hours. *Anesth Analg* 1991;73:530-5.
 43. Bernard J, Le Penven-Henninger C, Passuti N. Sudden decreases in mixed venous oxygen saturation during posterior spinal fusion. *Anesth*

- Analg 1995;80:1038-41.
44. Varin R, Mulder P, Tamion F, et al. Improvement of endothelial function by chronic angiotensin-converting enzyme inhibition in heart failure: role of nitric oxide, prostanoids, oxidant stress, and bradykinin. *Circulation* 2000;102:351-6.
 45. Auron M, Harte B, Kumar A, Michota F. Renin-angiotensin system antagonists in the perioperative setting: clinical consequences and recommendations for practice. *Postgrad Med J* 2011;87:472-81.
 46. Colson P, Ryckwaert F, Coriat P. Renin angiotensin system antagonists and anesthesia. *Anesth Analg* 1999;89:1143-55.
 47. Behnia R, Molteni A, Igic R. Angiotensin-converting enzyme inhibitors: mechanism of action and implications in anesthesia practice. *Curr Pharm Des* 2003;9:763-76.
 48. Coriat P, Richer C, Douraki T, et al. Influence of chronic angiotensin-converting enzyme inhibition on anesthetic induction. *Anesthesiology* 1994;81:299-307.
 49. Bertrand M, Godet G, Meersschaert K, et al. Should the angiotensin II antagonists be discontinued before surgery? *Anesth Analg* 2001;92:26-30.
 50. Comfere T, Sprung J, Kumar M, et al. Angiotensin system inhibitors in a general surgical population. *Anesth Analg* 2005;100:636-44.
 51. Wilson W, Taubert K, Gewitz M, et al. Prevention of infective endocarditis: guidelines from the American Heart Association: A guideline from the American Heart Association rheumatic fever, endocarditis, and Kawasaki disease committee, council on cardiovascular disease in the young, and the council on clinical cardiology, council on cardiovascular surgery and anesthesia, and the quality of care and outcomes research interdisciplinary working group. *Circulation* 2007;116:1736-54.
 52. Young B, Ng T, Teng C, et al. Non-concordance with surgical site infection prevention guidelines and rates of surgical site infections for general surgical, neurological, and orthopedic procedures. *Antimicrob Agents Chemother* 2011;55:4659-63.
 53. Deiner S. Highlights of anesthetic considerations for intraoperative neuromonitoring. *Semin Cardiothorac Vasc Anesth* 2010;14:51-3.
 54. Sloan T and Heyer E. Anesthesia for intraoperative neurophysiologic monitoring of the spinal cord. *J Clin Neurophysiol* 2002;19:430-43.
 55. Balvin M, Song K, Slimp J. Effects of anesthetic regimens and other confounding factors affecting the interpretation of motor evoked potentials during pediatric spine surgery. *Am J Electroneurodiagnostic Technol* 2010;50:219-44.
 56. Reinacher P, Priebe H, Blumrich W, et al. The effects of stimulation pattern and sevoflurane concentration on intraoperative motor-evoked potentials. *Anesth Analg* 2006;102:888-95.
 57. Lo Y, Dan Y, Tan Y, et al. Intra-operative monitoring in scoliosis surgery with multi-pulse cortical stimuli and desflurane anesthesia. *Spinal Cord* 2004;42:342-5.
 58. Tobias J. Strategies for minimizing blood loss in orthopedic surgery. *Semin Hematol* 2004;41:145-56.
 59. Carson J, Altman D, Duff A, et al. Risk of bacterial infection associated with allogenic blood transfusion among patients undergoing hip fracture repair. *Transfusion* 1999;39:694-700.
 60. Goodnough L, Brecher M, Kanter M, et al. Transfusion medicine: first of two parts - blood transfusion. *N Engl J Med* 1999;340:438-47.
 61. Parshuram C, Doyle J, Lau W, et al. Transfusion-associated graft versus host disease. *Pediatr Crit Care Med* 2002;3:57-62.
 62. Taylor R, Manganaro L, O'Brien J, et al. Impact of allogenic packed red blood cell transfusion on nosocomial infection rates in the critically ill patient. *Crit Care Med* 2002;30:2249-54.
 63. Florentino-Pineda I, Blakemore L, Thompson G, et al. The effect of epsilon aminocaproic acid on perioperative blood loss in patients with idiopathic scoliosis undergoing posterior spinal fusion. *Spine* 2001;26:1147-51.
 64. Schmeid H, Kurz A, Sessler D, et al. Mild hypothermia increases blood loss and transfusion requirements during total hip arthroplasty. *Lancet* 1996;347:289-92.