

## REVIEW ARTICLE

# Perioperative management following an atrial level switch (Mustard or Senning procedure) for transposition of the great arteries

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## 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. The current manuscript reviews the anatomy of TGA, outlines the past and current surgical approaches, and discusses the perioperative concerns of such patients.

**Key words:** Transposition of great arteries; Perioperative care; Mustard procedure; Senning procedure; Myocardial dysfunction; Congenital heart disease

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## INTRODUCTION

Congenital heart disease has a reported incidence of approximately 0.8 per 100 births.<sup>1,2</sup> 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. Currently alternative techniques are now available for the anatomic repair of patients with transposition of the great arteries (TGA); however, 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 review the anatomy of TGA, outline the past and current surgical approaches, and discuss the perioperative concerns of such patients.

## ANATOMY OF TRANSPOSITION & SURGICAL OPTIONS

Transposition of the great arteries (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.<sup>3</sup> 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 1:3,500-5,000 live births with a male:female ratio of 3.2:1.<sup>3</sup> 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.<sup>3</sup> As compared to a physiologically normal circulation system that exists in series, the parallel circuits that are present with D-TGA results in systemic cyanosis.

Post-natal 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).<sup>3</sup> 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.<sup>3</sup> 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.<sup>3</sup> 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.<sup>3</sup> Initially, 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.<sup>3</sup>

The Senning operation for TGA was first reported by Senning in 1959 with later work by Quaegebeur in the late 1970's.<sup>4,5</sup> 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.<sup>6,7</sup>

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.<sup>7</sup> 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 pantaloan-shaped patch" between the atria to create the baffle, again redirecting the blood flow in the same manner as the Senning procedure.<sup>7</sup> Owing to its simplified nature, the Mustard procedure quickly became the surgeon's operation of choice.

## LONG TERM OUTCOMES

Although consistent survival beyond the neonatal period was possible, neither the Senning nor the Mustard procedures were without long term complications. By the 1980's, troubling sequelae of atrial-level repairs including atrial arrhythmias, baffle leaks and obstructions, and right ventricular dysfunction were becoming increasingly evident. 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.<sup>8</sup>

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.<sup>6,7</sup> 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.<sup>6</sup> 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.<sup>6</sup> 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)<sup>6,8,9</sup> Kammeraad et al demonstrated that the presence of symptoms related to arrhythmias including palpitations, dizziness or syncope were associated with a 21.6-fold increase in the risk of a sudden death event.<sup>9</sup>

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.<sup>7</sup> Radiofrequency ablation is frequently utilized in patients with atrial tachyarrhythmias with a high success rate; however, the arrhythmia often recurs, necessitating placement of a permanent pacemaker. 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.<sup>10,11</sup> 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.<sup>9</sup> 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.<sup>6</sup> 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.<sup>12,13</sup> This differentiation in myocyte orientation and contractile motion normally allows the LV to function against high-resistance systems.<sup>13</sup>

An accompanying factor to RV dysfunction, which may further compromise myocardial performance, is the development of tricuspid valve regurgitation. In patients with atrial-level repairs, the anatomic tricuspid valve functions as the systemic atrioventricular valve and often becomes progressively incompetent.<sup>6</sup> Distention of the RV with progressive dysfunction and annular dilation may be one contributing factor; however, tricuspid regurgitation may precipitate the development of RV dysfunction as well.<sup>7</sup> The abnormal ventricular septum configuration may also play a contributing role. 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.<sup>6</sup> 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.<sup>6,7</sup> 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.<sup>6</sup> 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.<sup>6,7</sup> 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.<sup>14,15</sup>

Obstruction or leaking at the intra-atrial baffle is another, although infrequent, problem associated with the Senning and Mustard procedures. It is estimated that 5-15% of patients develop stenosis of the systemic venous baffle with a higher incidence following the Mustard repair.<sup>6</sup> The superior vena caval baffle limb is more frequently affected by obstruction and is often slowly progressive in nature. As a result of the insidious onset, true SVC syndrome is uncommon, owing to the development of venous collateral drainage. SVC baffle obstruction may be clinically silent, although patients can experience diminished exercise capacity as a result of the decreased venous return.<sup>6</sup> Unlike SVC baffle obstruction, IVC baffle obstruction is poorly tolerated and requires prompt evaluation with the relatively acute onset of symptoms related to the elevated venous pressure which leads to hepatic congestion, hepatomegaly and ascites.<sup>6,7</sup>

Small intra-atrial baffle leaks are more common than obstructions and frequently are not clinically apparent. When present, the leak usually functions as a left-to-right shunt; although hemodynamically insignificant, the presence of a shunt can place a patient at risk for a paradoxical embolus or cerebrovascular accident.<sup>7</sup> During the perioperative period, meticulous attention to avoid air is mandatory and the use of air filters is suggested when administering fluids and medications. In the setting of a large baffle leak, pulmonary and right atrial volume overload may become problematic with the development of congestive heart failure.<sup>6</sup> Lastly, depending on where the leak is located, the shunt may be from right-to-left, resulting in desaturation and cyanotic episodes with exercise.<sup>6</sup>

Owing to the development of several late complications from the Senning and Mustard procedures, an anatomic repair of TGA was sought and became feasible through the arterial switch procedure in the late 1980's. Originally

described by Jateen in 1976, the procedure involves transection of the aorta and pulmonary trunks above the sinuses, and separation of the coronary arteries from the pulmonary artery. The distal end of the pulmonary artery is placed anterior and connected to the right ventricle while the transected distal end of the aorta is relocated posteriorly to left ventricle. The coronary arteries are then translocated from the pulmonary artery to the aorta.<sup>2,7</sup> The development of this new procedure led to near abandonment of the Senning and Mustard procedures, in an attempt to minimize the late complications associated with those surgeries. The atrial level switches (Mustard and Senning) are occasionally performed today in patients with TGA and abnormal coronary artery anatomy which precludes their effective reimplantation and performance of the arterial switch procedure.

## **PERIOPERATIVE CARE**

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 2<sup>nd</sup> and 3<sup>rd</sup> decades 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
3. an evaluation of the baffle for obstruction or leaks

The initial evaluation of these patients should begin with a complete cardiac history, specifically questioning exercise capacity or intolerance, orthopnea or paroxysmal nocturnal dyspnea, the presence of palpitations, a documented history of arrhythmia, and prior syncopal events.<sup>6</sup> 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.

Patients who have undergone a Senning or Mustard repair frequently have abnormal electrocardiograms (ECG). 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 V<sub>1</sub> 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.<sup>6,7</sup> 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. Such complications include 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.<sup>16,17</sup> 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.<sup>16</sup> EMI may interfere with pacemaker function, which may be problematic if the patient is pacemaker-dependent for the majority of the time, or EMI may cause alterations in the programming of the device.<sup>17</sup> Current perioperative guidelines on cardiac rhythm management devices established by the American Society of Anesthesiologists (ASA) are outlined in reference 16. The recommendations include the following:

1. Establish that the patient has a device
2. Define the functionality of the device
3. Determine if the patient is pacemaker dependent
4. Confirm device function. The comprehensive evaluation of the device should be performed with an appropriate manufacturer-specific programming device by a knowledgeable consultant.

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. While the function



of the ICD is turned off, the patient should remain appropriately monitored, and external pads should be placed on the patient with an external defibrillatory source immediately available. The ASA further recommends that all devices be interrogated for the resumption of preoperative settings prior to discharge from the post-anesthesia care unit.<sup>16</sup>

For most modern devices, the continuous placement of a magnet over the pacemaker pulse generator will create an asynchronous mode, while magnet removal will result in reversion back to baseline device function. In the setting of an ICD, placement of a magnet over the device will disable the defibrillatory function; however the backup pacing function will remain active. When a combined pacemaker-ICD is present, use of a magnet will deactivate the ICD without changing the pacing function.<sup>16</sup> In all cases in which a magnet has been utilized, the ASA recommends postoperative device interrogation. Other precautions that can be employed to minimize EMI include placing the cautery grounding plate as distal as possible from the device; limiting cautery use to short, irregular bursts of cutting rather than coagulating current; and the use of bipolar cautery.<sup>16</sup>

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.<sup>18</sup> A thorough assessment of RV function and estimation of right ventricular ejection fraction using TTE can be difficult due to the complex shape of the RV and limited endocardial surface recognition as a result of heavy apical trabeculations.<sup>12</sup> 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.<sup>19</sup> In their study, Hoffman et al compared the right ventricular area change measured by TTE subcostal view to magnetic resonance perfusion imaging, and found that impaired systolic function detected with subcostal TTE predicted moderate-to-severe RV perfusion defects with 78% sensitivity and 62% specificity.<sup>19</sup> Further evaluation of RV function can be obtained with the use of either cardiopulmonary

exercise testing or dobutamine stress echocardiography. 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.<sup>20</sup> They also reported that systemic ventricular function is significantly depressed in up to 80% of patients post-atrial switch, and objective exercise capacity is impaired in these patients despite patient perception of normal or only mildly impaired function. 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.<sup>20,21</sup> In patients with poor TTE images, or those unable to undergo cardiopulmonary exercise testing, the use of TEE, cardiac magnetic resonance imaging (MRI), or cardiac catheterization may be necessary to fully evaluate systemic ventricular function, the presence of baffle leaks, and valvular function.

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.<sup>22</sup> 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.<sup>22</sup> If cardiac catheterization is performed, indicators of RV dysfunction or failure include a right atrial pressure (RAP) = 8-10 mmHg, a RAP to pulmonary capillary wedge pressure (PCWP) = 0.8, or a cardiac index = 2.2 L/min/M<sup>2</sup>.

Another diagnostic tool and prognostic marker that may be utilized to assess right ventricular function and volume status is the plasma concentration of B-type natriuretic peptide (BNP). 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).<sup>23,24</sup> Several studies, conducted in both the pediatric and adult population, have demonstrated a positive correlation between elevated BNP levels and the magnitude of systemic ventricular dysfunction. Knirsch et al demonstrated that right ventricular volume overload is more frequently associated with elevated BNP levels

when compared to left ventricular overload as volume overload in general is a more significant mechanism for BNP release than pressure overload.<sup>24</sup> Furthermore, a correlation between BNP levels and NYHA functional class/cardiopulmonary exercise capacity has been demonstrated with BNP levels greater than 45 pg/mL being predictive of significant right ventricular dysfunction.<sup>25</sup> 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.<sup>26</sup> 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.<sup>27</sup>

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.<sup>28,29</sup> Patients with concerning symptoms or a suspected history of PH should have further evaluation with an echocardiograph, which may demonstrate findings suggesting of PH including elevated RV systolic pressures (RVSP greater than 40 mmHg), right atrial enlargement, RV hypertrophy or interventricular septal flattening. When any of these findings are identified on echocardiography, further investigation with a right heart catheterization (RHC), which is the gold standard for evaluation of pulmonary hypertension, should be considered. Mean PAP greater than 25 mmHg, an increased PCWP, calculated pulmonary vascular resistance greater than 3 Woods units, and a normal or reduced cardiac output provide a definitive diagnosis of pulmonary hypertension. The level of perioperative risk in these patients is dependent on the degree of preoperative RV dysfunction and elevation in PVR, along with the type of surgical procedure. Any procedure that causes perioperative systemic inflammation, rapid blood loss, or has a high possibility of air, fat, carbon dioxide, or cement emboli would be considered high risk in this patient population. Several studies have demonstrated that children with PH have a significant risk of perioperative cardiac complications including cardiac arrest, pulmonary hypertensive crisis, and death.<sup>30,31</sup>

The perioperative management of patients with PH can be challenging. Patients on chronic therapy for PH

including intravenous prostacyclins (epoprostenol and trepostinil), phosphodiesterase inhibitors (sildenafil), endothelin antagonists (bosentan), and inhaled prostacyclin analogs (iloprost) should be continued on these medications throughout the perioperative period.<sup>30,31</sup> 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. A pulmonary hypertensive crisis is characterized by a rapid increase in PVR to the point that the PAP exceeds systemic blood pressure, RV ejection fraction acutely drops leading to a decrease in pulmonary blood flow, decreased cardiac output and biventricular failure.<sup>33</sup> 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 cmH<sub>2</sub>O) 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.

Despite appropriate measures to minimize elevations in PVR, pulmonary hypertensive crises may occur intraoperatively. Moderate hyperventilation with 100% oxygen, along with the initiation of inhaled nitric oxide (iNO), are the first line of treatments for intraoperative PH.<sup>32-34</sup> The benefit of iNO is typically achieved within 20 minutes of initiation and would be evidenced by improved oxygen saturations at lower inspired oxygen levels, along with a decrease in the ratio of PAP to systemic arterial pressure, increased cardiac index, decreased intrapulmonary shunting, and improved gas exchange. In addition to the use of iNO, the most important adjuvant therapy is aggressive alveolar recruitment to improve nitric oxide delivery.<sup>35</sup> Regardless of the dose administered, abrupt withdraw of iNO can result in severe rebound pulmonary hypertension and life-threatening hypoxemia. As a result, iNO should be continued postoperatively via endotracheal tube or face mask, weaned slowly with increasing oxygen

supplementation with consideration of the administration of oral or intravenous pulmonary vasodilators prior to discontinuation of iNO.

Senning and Mustard repair patients with both pulmonary hypertension and RV dysfunction may benefit from the perioperative use of milrinone. In patients with a low cardiac output and elevated mean PAP, milrinone increases cardiac output by augmenting contractility while decreasing PVR and SVR.<sup>35</sup> However, systemic hypotension as a result of vasodilation and reduced SVR remains the rate-limiting factor with milrinone use. 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  $\beta$ -adrenergic antagonists should be considered.<sup>36,37</sup>

RV preload should be optimized while avoiding volume overload, acute RV distention, and increased TR. Perioperative central venous pressure (CVP) monitoring is suggested to guide volume management in the setting of RV dysfunction. However, in the surgical setting with a patient placed in the prone position, CVP monitoring may be inaccurate. 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.<sup>38</sup> Based on simultaneous TEE imaging, these findings have been attributed to mechanical compression of the mediastinal structures, resulting in impaired ventricular filling and elevation of CVP. 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 may be warranted.

In addition to the use of TEE and CVP monitoring, continuous mixed-venous oxygen saturation (SvO<sub>2</sub>) may be used as an indicator of cardiac output and oxygen consumption.<sup>39,40</sup> The use of continuous SvO<sub>2</sub> monitoring can also serve as a guide to fluid and inotropic management with the demonstration that SvO<sub>2</sub>-guided therapy may reduce both hospital stay and postoperative complications.<sup>41</sup> A significant correlation between changes in cardiac output and measured SvO<sub>2</sub> has also been demonstrated in mechanically ventilated patients during anesthesia.<sup>42,43</sup>

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.<sup>44</sup> 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.<sup>45</sup> 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.<sup>46-48</sup> 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.<sup>46-48</sup> 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.<sup>49,50</sup> 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.

Prior to the initiation of surgery, it is also important to determine if the patient should receive bacterial endocarditis (SBE) prophylaxis. According to recent guidelines published by the American Heart Association, patients at high risk for SBE are those undergoing a dental, GI, or GU procedure and include patients with prosthetic cardiac valve or prosthetic material used for cardiac valve repair, patients with a previous history of infective endocarditis, cardiac transplant patients who develop valvulopathy, and CHD patients.<sup>51</sup> The category of CHD patients has been further clarified to include patients with unrepaired cyanotic CHD including palliative shunts and conduits, those with completely repaired CHD with prosthetic material within 6 months of the repair, and repaired CHD patients with residual defect at the site or adjacent to the site of prosthetic device or patch. Although many patients may not require antibiotics for SBE prophylaxis, the 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.<sup>52</sup> In the setting of major surgical procedures, unless contraindicated, either cefazolin or cefuroxime should be administered within 60 minutes before surgical incision. All prophylaxis should end within 24 hours after the operation and repeat dosing should occur if the operation lasts longer than 4 hours or if major blood loss occurs.



## SUMMARY

There are several potential perioperative considerations in caring for patients with TGA who have undergone an atrial level switch for TGA (Senning or Mustard procedure). Perioperative concerns include not only those related to the primary cardiac lesion, but also those related to the surgical procedure and its impact on intraoperative positioning and anesthetic requirements. 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. Preoperative evaluation will generally include echocardiography and consultation with cardiology to evaluate pacemaker/ICD function. Depending on the patient's cardiac status, the impact of anesthetic agents on myocardial function, sinus node function, and PVR should be considered.

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## My Most Unforgettable Experience

In my ancestral home there was one sparrow's nest in a wall crevice. Every year sparrow laid eggs and young chicks showed their beaks; their parents continually fed them. When they became old enough to fly they fled and never to return back to see their parents.

During my childhood and early adolescent days I used to think that everyone in this world was selfish, even perhaps the parents! They bring up their offspring so that they could get their help in old age. They are also selfish. If they did something for us, it's their job. But I was wrong; if our parents can help us to make it from trembling to strong feet they could take care of themselves in old age too.

When I became father I realized that what is being father. Whenever my daughter was hurt, I felt pain, as if I was hurt. When she became sick, I silently prayed that instead of her I might suffer. When she did something new we celebrated as she had had some invention. We spent several nights awake for our daughter but never complained; but can I do this for my parents or can she/he (my daughter or son) do the same for me? GOD knows better.

I don't remember whether I ever held my father's finger to learn walking (definitely I must have done but childhood memories do vanish fast), but I remember the day when he taught me to drive scooter. He sat as pillion rider and nervously, I left the clutch all of a sudden, the scooter jumped at 45 degrees. He fell down and must have hurt himself, but he didn't utter a word. He was my best teacher. It was the first professional exam during my medical graduation. The tension was so tremendous, I lost all the will to fight, the books seemed to bite back, and I lay limp in the bed sleepless. It was 2 am when I called him that I was nervous. He raced to my hostel which was 250 km away from my home and stood before me in 7 hrs before the start of my exam. Whenever I was in trouble I found him and his advice with me.

Today, he is suffering from frontal lobe infarct, an altered behavior and forgetfulness. I pay a visit once every 10 days even though my home is just 35 km away, since I remain busy in my job, my family and my patients. His hands tremor but my hands are nowhere to hold his. I earn at least thrice more than him still my father used to work as long as he could. Who is selfish, me or my father?

**Whatever we do for our parents we can't repay their debts.**

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The term congenitally corrected transposition of the great arteries describes a different entity that conjugates atrioventricular and ventriculoarterial discordance [2]. In the old nomenclature, completed transposition was used to describe either congenitally corrected or uncorrected transposition. In both situations, the great arteries were completely or predominantly misplaced across the ventricular septum. The arterial switch operation is the procedure of choice used to achieve complete physiological and anatomical repair. A repair at atrial level, either by a Mustard or a Senning procedure, is particularly suitable for hearts with an intact ventricular septum. The systemic venous return is redirected at atrial level to the left ventricle. Key words: Transposition of great arteries; Perioperative care; Mustard procedure; Senning procedure; Myocardial dysfunction; Congenital heart disease. Citation: Nicole Elsey N, Tobias JD. Perioperative management following an atrial level switch (Mustard or Senning procedure) for transposition of the great arteries (Review Article). *Anaesth Pain & Intensive Care* 2011;15(3):185-194. INTRODUCTION. Congenital heart disease has a reported incidence of approximately 0.8 per 100 births.<sup>1,2</sup> As such, patients with palliated or corrected congenital heart disease (CHD) may present for major surgical procedures. Two of these surgeries are the Mustard and the Senning repair (also called Mustard or Senning procedures). The main other surgeries are the arterial switch and Rastelli procedure. In this illustration of a heart with TGA that has had a Mustard/Senning repair, the blue blood flows through the two-way Systemic Venous Baffle (SVB) to the left ventricle. The blue blood is pumped to the lungs to get oxygen. Both are called "atrial switch procedures" because there is a baffle through the heart's top part, or atria, which allows the blood to reach the ventricles. When and where were Mustard and Senning repairs first done? Dr. Ake Senning of Sweden performed the first Senning procedure in 1957. Dr. William Mustard, from Canada, performed the first Mustard procedure in 1963. Since the mid-1980s, atrial switch procedures have been superseded by arterial switch procedures, which means that patients with Mustard or Senning repairs will disappear over the next 40 years [ 4 ]. In recent years, the atrial switch operation has been adopted only as part of the double switch operation for corrected transposition of the great arteries. Many patients who underwent the atrial switch operation survived to adulthood, even beyond 40 years of age. The survival rates of the Senning and Mustard procedures have been reported as similar.