Anaesthetic management and outcomes in patients with surgically corrected D-transposition of the great arteries undergoing non-cardiac surgery

R. E. Christensen*, P. I. Reynolds, B. K. Bukowski and S. Malviya

Department of Anesthesiology, F3900 C.S. Mott Hospital SPC 5211, The University of Michigan Health Systems, 1500 E. Medical Center Drive, Ann Arbor, MI 48109-5211, USA

*Corresponding author. E-mail: robertec@umich.edu

Background. Patients with effective repair of D-transposition of the great arteries (D-TGA) increasingly present for non-cardiac surgery. These patients may be predisposed to heart failure, arrhythmias, and sudden death, especially after the atrial switch repair. This retrospective study was undertaken to review the care and outcomes of patients with D-TGA who presented for non-cardiac surgery.

Methods. Records for patients with surgically corrected D-TGA undergoing general anaesthesia for non-cardiac surgery between October 2000 and April 2008 were reviewed. The anaesthesiology records, operative note, admission history and physical examination records, and discharge summaries of these patients were reviewed and the following data collected: patient characteristics; comorbidities; surgical procedure; anaesthetic and monitoring techniques; intra- and postoperative complications; and admission status.

Results. Fifty procedures, including 43 in the paediatric setting and seven in the adult setting, comprised the final sample. The majority of these patients received anaesthesia on an outpatient basis in the paediatric hospital, without invasive monitoring and without complication. There were four adverse events including a significant bradycardia, failed extubation after two of the procedures, and postoperative bleeding requiring return to the operating theatre in another.

Conclusions. Data suggest that the majority of patients with surgically corrected D-TGA can safely undergo general anaesthesia, often as outpatients, with no invasive monitoring. However, given the incidence of adverse events, it remains imperative that the perioperative care be individualized based on the presence of comorbidities, type of repair, residual cardiac disease, severity of planned surgery, and experience of the provider.

Keywords: complications; heart, congenital defects; surgery, non-cardiac; transposition of the great arteries

Accepted for publication: September 28, 2009

D-transposition of the great arteries (D-TGA) accounts for 4% of all congenital heart disease and is a defect in which there is atrioventricular concordance and discordance between the ventricles and the great arteries. The right ventricle gives rise to the aorta, whereas the left ventricle pumps blood to the pulmonary artery. With effective repair of D-TGA, the quality and duration of life for these patients have improved markedly.1 2 The long-term survival rates after surgical repair range from 76% to 96%, depending on the complexity of the defect and type of repair.3–5 Because of their increased survival, patients with surgically corrected D-TGA now present more frequently for non-cardiac surgery. Given their underlying pathophysiology, these patients may be at risk for heart failure, arrhythmias, and sudden death, particularly after the atrial switch repair.6 7 To date, however, very few data are available regarding the potential anaesthetic risks of this population. An understanding of the physiology and
complications of these patients is important to ensure their appropriate management for non-cardiac surgery. Indeed, a recent study found that 27% of perioperative cardiac arrests in children undergoing non-cardiac surgery occurred in those with congenital heart disease. This retrospective study was undertaken to review the anaesthetic care and outcomes of patients after complete repair of D-TGA who underwent non-cardiac surgery.

Methods
After Institutional Review Board approval, the perioperative electronic clinical information database (Centricity® General Electric Healthcare, Waukesha, WI, USA) was examined to identify all patients with a history of D-TGA who underwent general anaesthesia for a surgical or diagnostic procedure. This database contained the preoperative anaesthetic history, physical examination, and the anaesthetic records of all anaesthetics performed at the University of Michigan from October 2000 to April 2008. The following patients or procedures were excluded: those undergoing cardiac surgery, obstetric procedures, those with L-TGA, D-TGA associated with complex co-existing congenital heart defects (e.g. hypoplastic ventricle), and those undergoing interventional cardiology procedures. Patients who had a second surgery during the same hospitalization (e.g. tracheotomy for complications associated with the first procedure) were also excluded. Lastly, patients for which the repair method was unknown were excluded. This search yielded 52 procedures performed for 34 patients. The anaesthetic records were missing or incomplete in two procedures; these were excluded from the study, yielding a final sample of 50 procedures.

The anaesthesiology records, operative note, admission history and physical examination records, and discharge summaries of these patients were reviewed and the following data collected: patient characteristics; comorbidities; surgical procedure; anaesthetic and monitoring techniques; intra- and postoperative complications; and admission status. Data were summarized using descriptive statistics, and are presented as mean (SD) or n (%) as applicable.

Results
Fifty procedures, including 43 in the paediatric setting and seven in the adult setting, performed in 34 patients, comprised the final sample. Forty-five of these procedures were in patients who had undergone the arterial switch procedure for their initial repair, and five had undergone an atrial switch. A description of these procedures and the comorbidities of these patients are presented in Table 1.

In the majority of procedures, routine inhalation or i.v. agents were used for induction and maintenance of general anaesthesia with non-invasive monitoring (Table 2). Central venous pressure monitoring, pulmonary artery catheters, and transoesophageal echocardiograms were not used. Invasive arterial pressure monitoring was used in five procedures. Four of these were performed in the adult

Table 1 Description of the procedures (n=50). Data presented as n (%) or mean (range) as appropriate. *Residual heart defects= valvar or peripheral pulmonary stenosis, atrial or ventricular septal defects, aortic insufficiency, mitral regurgitation, and diminished right heart function. Pulmonary=mild reactive airway disease and one with severe restrictive lung disease secondary to scoliosis. Renal=insufficiency or structural abnormalities, none requiring dialysis. Neurologic=developmental delay, hydrocephalus and seizure disorders. Arrhythmia=complete heart block, sick sinus syndrome, pacemaker dependence and sinus brady cardiac episodes while sleeping. †Low severity surgery= non-invasive diagnostic or superficial surgery with minimal blood loss; moderate= invasive (anticipated moderate blood loss), emergent, or airway procedure; high= major procedure (anticipated excessive blood loss)
setting during surgeries with minimal expected blood loss. However, each of these patients had significant comorbidities, including mitral valve regurgitation, sick sinus syndrome, or diminished ventricular function. In three of these four procedures, patients had undergone the atrial switch (Mustard) repair. An arterial line was placed for only one procedure in the paediatric setting, an anterior spine fusion with significant expected blood loss.

The majority of patients tolerated the anaesthetic well and were discharged home or to a general care unit without adverse events. There were four adverse events (8%), including significant bradycardia during one procedure, failed tracheal extubation in two children, and postoperative bleeding requiring return to the operating theatre in another. The details of these procedures are described in Table 3.

### Table 3 Anaesthetic management. Data are given as n (%) 

<table>
<thead>
<tr>
<th>Event</th>
<th>Age (yr)</th>
<th>ASA</th>
<th>Procedure</th>
<th>Pertinent medical history</th>
<th>Intervention and outcomes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Severe bradycardia during insufflation of abdomen</td>
<td>15.6</td>
<td>III</td>
<td>Laparoscopic gynaecologic surgery</td>
<td>Sinus bradycardia and bundle branch block, pulmonary stenosis</td>
<td>Glycopyrrolate; sent to general care</td>
</tr>
<tr>
<td>Failed extubation</td>
<td>14.6</td>
<td>III</td>
<td>Anterior spine fusion</td>
<td>Mild pulmonary and tricuspid regurgitation, pulmonary stenosis; severe restrictive lung disease; dyspnoea on exertion and fatigue</td>
<td>Tracheotomy several days after surgery; remains ventilator-dependent 3 yr after operation</td>
</tr>
<tr>
<td>Failed extubation</td>
<td>0.25</td>
<td>III</td>
<td>Supraglottoplasty</td>
<td>Kabuki syndrome; Pierre Robin sequence, floppy epiglottis, laryngomalacia; mild aortic regurgitation and mild pulmonary artery stenosis</td>
<td>Tracheotomy; decannulated 1 yr later</td>
</tr>
<tr>
<td>Bleeding/haematoma</td>
<td>1.9</td>
<td>III</td>
<td>Circumcision</td>
<td>Mild pulmonary regurgitation and aortic insufficiency</td>
<td>Re-operation; discharged home after</td>
</tr>
</tbody>
</table>

### Discussion

This case series presents the largest review, to date, of patients with repaired D-TGA who underwent general anaesthesia for a non-cardiac procedure. Previous reviews have suggested that anaesthetic risk for patients with congenital heart disease undergoing non-cardiac procedures is dependent on the severity of the lesion, type of repair (complete vs palliative), status of the pulmonary vasculature, and ongoing comorbidities. Such reviews suggest that conventional anaesthetic techniques may be safely used in patients with congenital heart disease who had good surgical outcomes and no subsequent deterioration. Consideration of the type of repair and residual comorbidities is therefore imperative when providing anaesthetic care to patients with a history of D-TGA.

Indeed, the two types of surgical repair for this defect pose differing long-term outcomes. The atrial switch procedure (i.e. Mustard or Senning) involves redirection of blood flow from the vena cavae to the left ventricle, and pulmonary venous return to the right ventricle. After this procedure, the right ventricle pumps blood to the systemic circulation, placing a considerable strain on this thinned structure and posing a risk for right ventricular failure. Diminished right ventricular ejection fraction both at rest and during exercise has been reported in patients with D-TGA after the Mustard procedure. Patients who have undergone these procedures have a high incidence of ongoing arrhythmias (30–100%) because of damage to the sinoatrial or atrioventricular nodes or intra-atrial conduction pathways. Furthermore, these patients are at risk for obstruction to venous return and right ventricular failure. Only five patients in our sample had undergone atrial switch for repair of D-TGA. All of these patients had significant ongoing cardiac dysfunction, including heart block, sick sinus syndrome, and pulmonary stenosis. Anaesthetic management of these patients differed, in that all had an i.v. induction and the majority had invasive arterial pressure monitoring. However, despite the above risks, we found no adverse events in the five anaesthetics performed for atrial switch patients.

The atrial switch repairs were largely supplanted by the arterial switch (Jatene procedure) in the mid-1980s, wherein blood flow is redirected at the level of the arteries. The aorta and pulmonary arteries are switched so that the left ventricle supports the systemic circulation and the right ventricle provides pulmonary blood flow. This procedure avoids the development of right ventricular failure; however, neo-aortic root dilation and neo-aortic regurgitation have been reported to occur during follow-up.
Although the arterial switch procedure reduced the incidence of rhythm disturbance after operation, a 20–30% incidence of supraventricular and ventricular ectopy have been reported. Additionally, long-term effects of this repair on the coronary arteries and systemic circulation remain unknown. A significant number of patients in this series who had undergone arterial switch had residual cardiovascular and other comorbidities upon presentation for non-cardiac surgery. However, routine anaesthetics and non-invasive monitoring were sufficient to ensure safe care and outcomes in the majority of these patients. Importantly, 8% of these patients had an adverse event during or immediately after surgery. However, only one of these events was likely related to the underlying residual cardiac condition (i.e. bradycardia). Two were related to significant pulmonary or airway comorbidities. The first of these patients presented with a history of severe restrictive lung disease secondary to scoliosis. Poor pulmonary function before operation likely led to the need for long-term ventilatory support. In the second, postoperative airway obstruction and the need for tracheotomy were likely related to pre-existing laryngomalacia. Both patients required intensive care unit management after operation. The last complication was one of surgical haemostasis. The importance of vigilance in monitoring with consideration of the surgical and patient risk factors, which may include the potential for arrhythmias, heart failure, or both, cannot be over-emphasized for the safe care of patients with D-TGA.

This study is subject to the limitations of any retrospective study. Patients who met inclusion criteria could have been missed by our search of the computerized database. Additionally, because of the retrospective study design, some of the minor complications may have been missed. The sample included subsets of patients with small numbers such as those who had undergone atrial switch repairs and adult patients, making it difficult to describe the perioperative risks in these groups. Lastly, this study was conducted at a tertiary centre with extensive experience with congenital heart disease, which may hinder the ability to generalize findings to other settings.

This study describes the anaesthetic experience in patients with previous repair of D-TGA, a rare and complex heart defect. A variety of anaesthetic techniques were successfully used, often on an outpatient basis without invasive monitoring. Adverse events were, in general, related to underlying comorbidities. However, given the significant incidence of adverse events, it remains imperative that each such case be carefully evaluated and the perioperative care be individualized based on the presence of comorbidities, type of repair, residual cardiac disease, severity of planned surgery, and experience of the provider.

Acknowledgement
The authors would like to acknowledge the invaluable aid of Terri Voepel-Lewis in the research and preparation of this manuscript.

References