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Anaesthesia for non-cardiac surgery in children and young adults with Fontan physiology

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Abstract

Introduction: Patients with Fontan physiology require non-cardiac surgery. Our objectives were to characterise perioperative outcomes of patients with Fontan physiology undergoing non-cardiac surgery and to identify characteristics which predict discharge on the same day. *Materials and Method:* Children and young adults with Fontan physiology who underwent a non-cardiac surgery or an imaging study under anaesthesia between 2013 and 2019 at a single-centre academic children's hospital were reviewed in a retrospective observational study. Continuous variables were compared using the Wilcoxon rank sum test, and categorical variables were analysed using the Chi-square test or Fisher's exact test. Multivariable logistic regression analysis results are presented by adjusted odds ratios with 95% confidence intervals and p values. *Results:* 182 patients underwent 344 non-cardiac procedures with anaesthesia. The median age was 11 years (IQR 5.2–18), 56.4% were male. General anaesthesia was administered in 289 (84%). 125 patients (36.3%) were discharged on the same day. On multivariable analysis, independent predictors that reduced the odds of same-day discharge included the chronic condition index (OR 0.91 per additional chronic condition, 95% CI 0.76–0.98, p = 0.022), undergoing a major surgical procedure (OR 0.17, 95% CI 0.05–0.64, p = 0.009), the use of intraoperative inotropes (OR 0.48, 95% CI 0.25–0.94, p = 0.031), and preoperative admission (OR = 0.24, 95% CI: 0.1–0.57, p = 0.001). *Discussion:* In a contemporary cohort of paediatric and young adults with Fontan physiology, 36.3% were able to be discharged on the same day of their non-cardiac procedure. Well selected patients with Fontan physiology can undergo anaesthesia without complications and be discharged same day.

Patients with multiple types of CHD may be palliated with a Fontan procedure. The Fontan procedure is suitable for patients who are not amenable to a two-ventricle repair due to either a small or absent ventricle, or due to complex anatomy which makes septation difficult.[1–3] In a recent publication from the Pediatric Heart Network, Fontan patients were found to have a 95% transplant-free survival at an average of 7 years of follow-up.[4] Many patients with Fontan physiology will live for decades[5,6] and thus may require diagnostic imaging and surgical procedures. Importantly, many of these interventions will require general anaesthesia or sedation.

Despite the large numbers of patients with Fontan circulations, analysis of the periprocedural outcomes following non-cardiac procedures has been limited. Our objectives are to describe the periprocedural course for non-cardiac procedures for patients with Fontan physiology and identify the predictors for discharge home the same day.

Methods

Institutional review board approval was obtained (IRB-P00022312). Administrative databases were interrogated for the interval between 1 January, 2013 and 31 December, 2019 to identify all patients who had undergone a previous Fontan procedure (n = 871). Anesthesia databases were then interrogated to identify any anaesthetic encounters for the cohort of Fontan patients. Anesthetics for cardiac surgery, cardiac catheterisation, electrophysiology or arrhythmia procedures, and cardiac imaging were all excluded. Anesthetics were excluded if they occurred either prior to the Fontan procedure (i.e. at the time of bidirectional Glenn) or if the patient no longer had a Fontan circulation following heart transplant, conversion to biventricular circulation, or had undergone a Fontan take-down to another single-ventricle palliation. An additional four anaesthetics were removed, because no medications were administered by an anaesthesiologist. This resulted in a final cohort of 182 patients who underwent 344 non-cardiac procedures under the care of a paediatric anaesthesiologist.

Data were collected using chart review and electronic capture of data from the electronic medical record to include basic demographics, underlying cardiac disease, type of Fontan, ventricular function and presence of valvular dysfunction on most recent echocardiogram, and baseline oxygen saturation. Details recorded from the anaesthesia record included the start time, the length of the anaesthetic, type of anaesthesia (general versus monitored anaesthesia care), airway management, use of neuromuscular blocking agents, use of inotropes, and the volumes of crystalloid and packed red blood cells administered. Comorbid conditions were assessed using a chronic condition indicator. Chronic condition indicator is an 18-category tool developed to classify International Classification of Diseases (ICD)-10 codes as chronic within administrative databases (Healthcare Cost and Utilization Project – HCUP).[7] A chronic condition is defined as one which is present for 12 months or greater, requires ongoing medical care, or results in a limitation of self-functioning. Patients were assigned 1 point for each system other than cardiovascular which was affected by 1 or more chronic conditions. The final disposition of the patients was determined based on the final location of recovery.

Procedures were categorised as diagnostic imaging, minor surgical procedures, or major surgical procedures. Minor procedures were defined as those which could reasonably be expected to be done with minimal sedation in a healthy, cooperative adults such as dental procedures, Botox injections, peripherally inserted central catheters, chest tubes, and liver biopsies. Also included in this category were direct laryngoscopy or endoscopy and dental rehabilitation. Major procedures included procedures with the potential for bleeding or major haemodynamic consequences such as most orthopaedic, neurosurgical, and general surgical procedures. Major procedures would be expected to require general anaesthesia.

Cardiopulmonary adverse events in patients included arrhythmias or tachy/bradycardias requiring intervention, increased oxygen requirement postoperatively to achieve baseline oxygen saturations after transfer to the recovery room, change to a more invasive airway (natural airway to laryngeal mask or endotracheal tube), cardiopulmonary resuscitation, or need for extracorporeal membrane oxygenation. These complications were assessed for the first 24 hours after anaesthesia and were only assessed for patients who did not recover in an ICU.

Statistics

Continuous data are reported as means and standard deviations, and categorical data are presented as frequencies and percentages. Continuous variables were compared using the Wilcoxon rank sum test, and categorical variables were analysed using the Chi-square test or Fisher's exact test. A two-tailed $p < 0.05$ was considered statistically significant. Multivariable logistic regression analysis results are presented by adjusted odds ratios with 95% confidence intervals and p values. A two-tailed alpha level of 0.05 was used to determine statistical significance. Stata (version 16.1, StataCorp LLC., College Station, Texas) was used for all statistical computing.

Results

We identified 182 patients who had 344 non-cardiac procedures between 1 January, 2013 and 31 December, 2019. The median number of anaesthetics per patients was 2 (IQR 1–5). The median age was 11 years (IQR 5.2–18.0), the median weight was 31.5 kg (18–54.6), and 194 (56.4%) were male. The ASA category was II in 14 (4.1%), III in 221 (64.2%), IV in 108 (31.4%), and V in 1 (0.3%) and were emergent procedures in 39 (11.3%).

Patient underlying diagnoses included hypoplastic left heart syndrome (n = 134, 39.0%), tricuspid atresia (n = 59, 17.2%), double-outlet right ventricle (n = 39, 11.3%), pulmonary atresia (n = 35, 10.2%), double-inlet left ventricle (n = 22, 6.4%), unbalanced complete atrioventricular canal (n = 17, 4.9%), and other diagnoses (n = 38, 11.0%). It was a systemic left ventricle in 177 patients (51.5%). Two hundred and sixty-seven (77.6%) patients had a lateral tunnel or intracardiac Fontan and 77 (22.4%) had an extracardiac Fontan. No patients in this series had a classical Fontan. Three hundred twenty-one patients (93.3%) had a preoperative echocardiogram within the prior year. Twenty-seven patients (8.4%) had a significant lesion burden as determined by echocardiography and defined as severe atrioventricular valve regurgitation (n = 8) and/or qualitatively severe dysfunction of the systemic ventricle (n = 20).

The median value for the chronic condition indicator was 3 (IQR 0–5). The most common non-cardiac diagnoses were in the digestive system (39.8%). Of these, the 2 most common coded diagnoses were gastroesophageal reflux disease and gastrostomy. Psychological conditions were present in 38.4% including most frequently acute stress reaction and generalised anxiety disorder. Respiratory conditions were present in 23.8% with the most common diagnoses being asthma, obstructive sleep apnoea, and oxygen dependency.

Anesthetic and surgical information are included in Table 1. General anaesthesia was used in 289 (84.0%) of anaesthetic encounters. Induction techniques included inhalational in 87 (25.3%), intravenous 247 (n = 71.8%), or a combination in 10 (2.9%). Airway management was a tracheostomy in 3 (0.9%), an endotracheal tube in 215 (62.5%), a laryngeal mask or mask in 33 (9.6%), or a natural airway in 93 (27.0%) of anaesthetics. Invasive monitoring included an arterial line in 48 (14.0%) anaesthetics and a central line or peripherally inserted central catheter in 44 (12.8%). Inotropes/vasopressors were used in 134 anaesthetics (39.0%) and included dopamine (n = 110, 32.0%), ephedrine (n = 36, 10.5%), epinephrine (n = 3, 0.9%), and calcium gluconate (n = 24, 7.0%). Neuromuscular blockade was used in 164 anaesthetics (47.8%). Patients received a median of 7.2 mL/kg/hour of crystalloid (IQR 3.7–10.2 mL/kg/hour). Packed red blood cells were administered in 16 anaesthetics (4.7%).

There was a total of 20 regional blocks in 16 patients undergoing general anaesthesia. This included a transversus abdominis plane or rectus sheath block (n = 8), supraclavicular or infraclavicular (n = 3), lower limb (femoral, facia iliaca, and adductor canal) (n = 4), lumbar plexus (n = 3), paravertebral (n = 1), and epidural (n = 1).

In 274 cases (79.7%), the patients were transferred to a post-anesthesia care unit. The median length of stay in the post-anesthesia care unit was 98 minutes (IQR 67–157). The median day admitted for all patients postoperatively was 1 day (IQR 0–7). One hundred twenty-five (36.3%) were discharged on the same day, 156 (45.3%) were admitted for a median of 2 days (IQR 1–11 days), and 63 (18.3%) went to the ICU. All patients who were discharged home the same day had a follow-up phone call by our nursing staff. There were no readmissions within 24 hours after discharge.

There were 61 patients who initially recovered in an ICU postoperatively as scheduled and 2 cases patients initially admitted to the post-anesthesia care unit with a subsequent need for transfer to an ICU due to lethargy with hypotension. Cardiorespiratory adverse events occurred during an additional 16 anaesthetics; 2 were cardiac (1 tachyarrhythmias and 1 bradyarrhythmia) and 14 were respiratory (new oxygen requirements above baseline in the recovery room (n = 9), unanticipated changes in airway

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Table 1. Anesthesia and surgery details.

Anesthetic and surgical details	n = 344
Location of anesthesia	
Main operating room	267 (77.6%)
Interventional radiology	50 (14.5%)
Radiology (MRI or CT scan)	27 (7.9%)
Time of day anaesthesia started	
Started between 7:00 and 13:00	226 (65.7%)
Started between 13:00 and 19:00	110 (32.0%)
Started between 19:00 and 7:00	8 (2.3%)
Median anaesthesia duration (mins)	102.5 (70.3–167.8)
Types of surgery or imaging	
Dental/oral maxillary	90 (26.2%)
Interventional radiology procedures	50 (14.5%)
Airway procedures	33 (9.6%)
Plastics/soft tissue	32 (9.3%)
Endoscopy	31 (9.0%)
Imaging	27 (7.8%)
Orthopedics	21 (6.1%)
Spine	12 (3.5%)
Open abdominal	12 (3.5%)
Botox/pain/bone marrow	10 (2.9%)
Laparoscopic	8 (2.3%)
Ophthalmology	6 (1.7%)
Neurosurgery	6 (1.7%)
Genitourinary	5 (1.5%)
Thoracic	1 (0.3%)
Classification of surgery types	
Diagnostic imaging	27 (7.8%)
Minor procedure	230 (66.9%)
Major procedure	87 (25.3%)
General anaesthesia	289 (84.0%)
Induction	
Intravenous	247 (71.8%)
Inhalational	87 (25.3%)
Combination	10 (2.9%)
Airway management	
Tracheostomy	3 (0.9%)
Endotracheal tube	215 (62.5%)
Laryngeal mask airway (LMA) or mask	33 (9.6%)
Natural airway	93 (27.0%)
Arterial line	48 (14.0%)
Central line or Peripherally Inserted Central Catheter (PICC)	44 (12.8%)
Any inotropes	134 (39.0%)
Neuromuscular blockade used	164 (47.8%)
Crystalloid (ml/kg per hour)	7.2 (3.7–10.2)
Packed red blood cells	16 (4.7%)

management intraoperatively (n = 3), laryngospasm (n = 1), and postoperative pleural effusion (n = 1)). There were no instances of cardiac arrest, respiratory arrest, extracorporeal membrane oxygenation initiation, or death within 24 hours. There were no complications related to regional anaesthesia in this cohort.

Univariate predictors of discharge on the same day are listed in Table 2. On univariate analysis, the following were associated with discharge: female gender ($p < 0.001$), ASA II or III ($p < 0.001$), elective procedures ($p < 0.001$), baseline oxygen saturations > 90% ($p < 0.001$), lower chronic condition index ($p < 0.001$), preoperative admission ($p < 0.001$), location of procedure ($p < 0.001$), anaesthesia duration ($p < 0.001$), anaesthesia start time ($p < 0.001$), type of procedure ($p < 0.001$), general anaesthesia ($p = 0.033$), type of airway device ($p = 0.019$), absence of arterial line ($p < 0.001$), absence of central venous access ($p < 0.001$), no inhaled anaesthetics used ($p = 0.027$), induction technique ($p < 0.001$), absence of inotropes used ($p < 0.001$), volume of crystalloid given per kg per hour ($p < 0.001$), and no blood transfusion ($p < 0.001$). On multivariable analysis, independent predictors that reduced the odds of same-day discharge included the chronic condition index (OR 0.91 per additional chronic condition, 95% CI 0.76–0.98, $p = 0.022$), undergoing a major surgical procedure (OR 0.17, 95% CI 0.05–0.64, $p = 0.009$), the use of intraoperative inotropes (OR 0.48, 95% CI 0.25–0.94, $p = 0.031$), and preoperative admission (OR = 0.24, 95% CI: 0.1–0.57, $p = 0.001$) (Table 3).

Discussion

Analysis of outcomes in Fontan patients undergoing non-cardiac procedures has been limited to adult patients[8,9] or patients undergoing high-risk surgery such as spine surgery.[10,11] We present an analysis of the largest series of children and young adults with a Fontan circulation undergoing a large variety of non-cardiac procedures. In our cohort, in 125 (36.3%) of anaesthetic, patients were successfully discharged the same day without readmission at 24 hours.

A lower number of chronic medical conditions was associated with same-day discharge (OR 0.91 per additional chronic condition, 95% CI 0.76–0.98, $p = 0.022$). The contribution of medical disease is clearly important. A recent study showed that patients with CHD and at least 1 chronic condition had a greater length of stay (21 days; IQR, 5–68) than those with CHD only (9 days; IQR, 3–46). End-organ dysfunction is common among patients with Fontan physiology, and this can play an important role in both drug metabolism and overall rate of recovery.

The use of inotropes was an independent predictor of need for admission, while the amount of crystalloid received was not associated in the multivariable model. While echocardiography can identify systolic dysfunction, we did not have enough information to assess the diastolic dysfunction which can be challenging in single-ventricle patients.[13] The use of inotropes is a decision made by the anaesthesia team in order to maintain systemic blood pressures. As such, it may be a surrogate for overall poor circulatory

Table 2. Univariate associations with same-day discharge.

Variable	Same-day discharge n = 125	Not discharged n = 219	p-Value
Age (years)	11 (5.8, 18)	11 (5, 18)	0.483
Weight (kg)	31.7 (19.2, 57.7)	31.3 (15.8, 50.9)	0.098
Female	86 (68.8%)	108 (49.3%)	**<0.001**
ASA category IV or V	15 (12%)	93 (42.5%)	**<0.001**
Emergent or urgent	1 (0.8%)	38 (17.4%)	**<0.001**
Baseline oxygen saturations			**<0.001**
90–100%	116 (92.8%)	156 (71.2%)	
<90%	9 (7.2%)	63 (28.8%)	
Left dominant ventricle	63 (50.4%)	114 (52.1%)	0.38
High risk feature on preoperative echocardiogram	6/109 (5.5%)	21/212 (9.9%)	0.208
Chronic condition index	1 (0, 3)	3 (1, 6)	**<0.001**
Location			**0.001**
Main operating room	109 (87.2%)	158 (72.2%)	
Out of operating room	16 (12.8%)	61 (27.9%)	
Anesthesia duration (minutes)	86 (64, 157)	112 (76, 185)	**<0.001**
Time of day			**<0.001**
Started before 13:00	104 (83.2%)	122 (55.7%)	
Started before 19:00	20 (16%)	90 (41.1%)	
Evening or night	1 (0.8%)	7 (3.2%)	
Admitted preoperatively	12 (9.6%)	125 (57.1%)	**<0.001**
Types of procedure			**<0.001**
Diagnostic imaging	10 (8%)	17 (7.8%)	
Minor	99 (79.2%)	131 (59.8%)	
Major	16 (12.8%)	71 (32.4%)	
General anaesthesia	112 (89.6%)	177 (80.8%)	**0.033**
Airway Endotracheal tube	79 (63.2%)	136 (62.1%)	**0.019**
Laryngeal Mask Airway (LMA) or mask	19 (15.2%)	14 (6.4%)	
Natural airway	26 (20.8%)	67 (30.6%)	
Tracheostomy	1 (0.8%)	2 (0.9%)	
Arterial line	1 (0.8%)	47 (21.5%)	**<0.001**
Central line or PICC	0 (0%)	44 (20.1%)	**<0.001**
Induction technique			**<0.001**
IV	73 (58.4%)	174 (79.5%)	
Inhaled	47 (37.6%)	40 (18.3%)	
Combination	5 (4%)	5 (2.3%)	
Inhaled anaesthetics used	103 (82.4%)	156 (71.2%)	**0.027**
Neuromuscular blockade used	56 (44.8%)	108 (49.3%)	0.42
Inotropes used intraoperatively	33 (26.4%)	101 (46.1%)	**<0.001**
Crystalloid volume per kg per hour	9 (6.5, 13)	5.6 (2.8, 9.2)	**<0.001**
Packed red blood cells	0 (0%)	16 (7.3%)	**0.001**

Continuous data are presented as median (IQR) and categorical data are presented as n (%).
Continuous variables were compared using the Wilcoxon rank sum test, and categorical variables were analysed using the Chi-square test or Fisher's exact test.

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Table 3. Multivariable logistic regression of same-day discharge.

Covariate	Adjusted odds ratio	95% CI	p-Value
Emergent or urgent	0.17	(0.02, 1.56)	0.118
Baseline oxygen saturations			
90–100%	Reference	.	.
<90%	0.46	(0.18, 1.16)	0.101
High risk feature on preoperative echocardiogram	0.6	(0.17, 2.04)	0.412
Chronic condition index (per additional chronic condition)	0.86	(0.76, 0.98)	**0.022***
Anesthesia duration (per 30 minutes)	0.91	(0.81, 1.02)	0.107
Time of day			
Started before 13:00	Reference	.	.
Started before 19:00	0.33	(0.15, 0.71)	**0.004***
Evening or night	1.68	(0.12, 22.8)	0.699
Admitted preoperatively	0.24	(0.1, 0.57)	**0.001***
Types of procedure			
Diagnostic imaging	Reference	.	.
Minor	0.76	(0.24, 2.38)	0.64
Major	0.17	(0.05, 0.64)	**0.009***
Airway			
Endotracheal tube	Reference	.	.
Laryngeal Mask Airway (LMA) or mask	1.95	(0.62, 3.46)	0.251
Natural airway	0.75	(0.28, 1.97)	0.554
Tracheostomy	0.32	(0.02, 4.86)	0.413
Neuromuscular blockade used	1.46	(0.62, 3.45)	0.384
Inotropes used intraoperatively	0.48	(0.25, 0.94)	**0.031***
Crystalloid volume per kg per hour	1.05	(0.98, 1.12)	0.146

*Statistically significant.

status. It is also likely that the need for inotropes would suggest to the treating clinicians that they are less likely to be successfully discharged home.

Patients who underwent major procedures were more likely to be admitted postoperatively, but this would be expected of patients without significant cardiac comorbidities as well. Interestingly, we did not find any association with the type of airway technique employed, the use of neuromuscular blockade, or the anaesthesia duration to be associated with hospital discharge. This supports the contention that the best technique for anaesthetizing a patient with Fontan circulation is unlikely to be a "one size fits all" strategy but rather a technique tailored to the patient's current physiologic condition, and the procedure being performed.

Due to differences in patient demographics, it is difficult to compare our results to other published series. In a study from the Mayo Clinic, there were 39 anaesthetics that were delivered with a 31% complication rate and 1 postoperative death.[8] However, the Fontan patients in this study were all adults of which 61.3% had an atriopulmonary connection, a form of cavopulmonary connection known to be associated with increased morbidity and mortality independent of non-cardiac surgery encounters. A larger follow-up study from the Mayo Clinic of 154 adult Fontan patients found a 12% intraoperative complication rate and a 5% postoperative complication rate including one death (0.2%).[9] In these adult patients, however, only 13% underwent a general anaesthetic which is very different from our population of children and young adults with Fontan physiology. In a small study of 36 patients with hypoplastic left heart syndrome palliated to a Fontan undergoing non-cardiac surgery, adverse events consisted of respiratory instability in 3 (8%) and airway obstruction or laryngospasm in 2 (6%) patients. Of note, there were no cases of cardiovascular instability.

We do not have strict institutional guidelines for same-day discharge, hospital admission, or ICU admission but rather rely on clinical judgement of the primary cardiologist, the consulting cardiac anaesthesiologist, and the primary anaesthesiology team. Patients with cardiovascular disease who undergo non-cardiac surgery or imaging at our institution are cared for by paediatric anaesthesiologists in the general operating rooms. While cardiac anaesthesiologists provide consultative service during the preoperative phase including medical optimisation, intraoperative management strategies, and postoperative disposition, it is the primary paediatric anaesthesia team that assumes responsibility for the care of these patients intraoperatively and in the recovery room.

Study limitations include all those inherent to a retrospective review. The decision process which determines patient disposition is difficult to ascertain in a retrospective study. Of note, patients at our institution are generally anticoagulated with aspirin alone and thus do not require admission for restarting anticoagulation. Specific information about anticoagulation was not collected. While this is a single-centre study which may limit generalizability, it is a large study that provides granular data unavailable in current registries and administrative databases. Because patients with Fontan procedures are not routinely catheterised immediately prior to non-cardiac surgery or imaging, no cardiac catheterisation data were collected or analysed.

Patients with Fontan physiology can successfully undergo same-day surgery under anaesthesia. The type of anaesthetic was not associated with discharge home. However, the presence of chronic medical conditions, undergoing a major surgical procedure, preoperative admission, and need of inotropic support during the procedure are associated with reduced odds of same-day discharge.

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Conflict of Interest. None.

Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008, and have been approved by the institutional committees (Boston Children's Hospital Institutional Review Board).

References

1. Gewillig M, Brown SC. The Fontan circulation after 45 years: update in physiology. Heart 2016; 102: 1081–1086.
2. Yuki K, Al Casta, Uezono S. Anesthetic management of noncardiac surgery for patients with single ventricle physiology. J Anesth 2011; 25: 247–256.

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3. Jolley M, Colan SD, Rhodes J, DiNardo J. Fontan physiology revisited. Anesth Analg 2015; 121: 172–182.
4. Atz AM, Zak V, Mahony et al. Survival data and predictors of functional outcome an average of 15 years after the Fontan procedure: the Pediatric Heart Network Fontan Cohort. Congenit Heart Dis 2015; 10: E30–42.
5. Ono M, Boethig D, Goerler H, et al. Clinical outcome of patients 20 years after Fontan operation-effect of fenestration on late morbidity. Eur J Cardiothorac Surg 2006; 30: 923–929.
6. Reddy S, Siehr Handler S, Wu S, Rabinovitch M, Wright G. Proceedings from the 2019 Stanford single ventricle scientific summit: advancing science for single ventricle patients: from discovery to clinical applications. JAHA 2020; 9: e015871.
7. Agency for Healthcare Research and Quality, Healthcare Cost and Utilization Project. User guide: chronic condition indicator for ICD-10-CM, v2021.1 (Beta version) 2020. Accessed May 24, 2021. *Available at:* https://www.hcup-us.ahrq.gov/toolssoftware/chronic_icd10/chronic_icd10.jsp.
8. Rabbitts JA, Groenewald CB, Mauermann WJ, et al. Outcomes of general anesthesia for noncardiac surgery in a series of patients with fontan palliation. Pediatr Anaesth. 2013; 23: 180–187.
9. Egbe AC, Khan AR, Ammash NM, et al. Predictors of procedural complications in adult Fontan patients undergoing non-cardiac procedures. Heart. 2017; 103: 1813–1820.
10. Uehara M, Takahashi J, Akazawa Y, et al. Posterior spinal fusion for scoliosis after Fontan procedure. J Orthoped Sci. 2018; 23: 294–298.
11. Przybylski R, Hedequist DJ, Nasr VG, et al. Adverse perioperative events in children with complex congenital heart disease undergoing operative scoliosis repair in the contemporary era. Pediatr Cardiol. 2019; 40: 1468–1475.
12. Bundts W, Ravekes WJ, Danford DA, Kutty S. Diastolic heart failure in patients with the Fontan circulation: a review. JAMA Cardiol. 2020; 5: 590–597.
13. D'Udekem Y, Iyengar AJ, Cochrane AD, et al. Contemporary techniques have improved long-term outcomes. Circulation 2007; 116: I157–I164.