A case series of different anesthesia approaches for single ventricular physiology patients in various stages of palliation underwent noncardiac procedures

ABSTRACT
Patients with single ventricle physiology (SVP) are a particularly challenging population with congenital heart disease (CHD); they will go for staged, palliation ending in the Fontan circulation. Nowadays, with improvement in surgical procedures for CHD, these patients become growing population, and noncardiac surgeries become not uncommon. The authors report different anesthesia approaches for four pediatric patients with SVP underwent ten noncardiac procedures done under general anesthesia following the different stages of palliation at King Abdulaziz Medical City, Riyadh, Kingdom of Saudi Arabia, between 2009 and 2015 and do a brief review of the literature on this topic. The aim of this study is to highlight that anesthesia approach for patients with SVP varies according to the patient physical situation at the time of the procedure, stage of palliation, and type of surgery. Therefore, every anesthesiologist should have thorough knowledge about SVP, different stages of palliative surgery, anesthesia concern in each one and risk factors associated with perioperative morbidity before anesthetizing patients for a noncardiac procedure to keep patient safety as well as avoiding unnecessary cancellation, rescheduling, and admissions to the ward or the Intensive Care Unit.

Key words: Anesthesia of noncardiac surgery; congenital heart disease; Fontan procedure; Glenn circulation; intraoperative complications; postoperative complications; risk factors; single ventricle physiology

Introduction
Congenital heart disease (CHD) is the most common congenital anomalies, occurring in approximately 4–9 of 1000 live births.[1] Nowadays, with improvement in surgical procedures for CHD, these patients become growing population, and noncardiac surgeries become not uncommon. Single ventricle patients are a challenging group from CHD who will go for staged palliation ending in the Fontan circulation, and some studies described them as high risk.[2]

Different studies identify a variety of factors associated with a high risk of perioperative complications in patients with CHD for noncardiac surgery, of which is how complicated are the disease, the general and physiological condition, type...
of operation, and the smaller the child. The most important factors are the general condition, and how complicated is the disease, what sort of corrective cardiac surgery, is it complete or only palliative.[3]

Those patients with CHD who have the good general condition can proceed for elective procedures with a low risk while those who are not well compensated and going for emergency or major operations are at high risk.[3]

This study describes the perioperative management of several pediatric patients with different stages of palliation for single ventricle physiology (SVP) undergoing general anesthesia for noncardiac surgery at a tertiary care institution within the period of 2009–2015.

It will also highlight the need for different anesthetic approaches for management for these patients according to the stage of palliation, present of risk factors, and type of procedures.

**Case Reports**

**Case 1**
The first patient was a female who had tricuspid atresia, dextro-transposition of great arteries, ventricular septal defect, and hypoplastic right ventricular (RV) as primary CHD; first, she had BT shunt as the first step in palliative procedures, her problem was the elevation of pulmonary artery pressure, she had hemodynamic assessment in cardiac catheterization laboratory which was not in favor of Glenn, and hence, the decision was to do pulmonary artery banding with atrial septectomy and keep the patient medicated with furosemide and sildenafil. Before proceeding to banding, she had standard American Society of Anesthesiologists (ASA) monitors applied and induction of anesthesia using ketamine 2 mg/kg and fentanyl 2 mcg/kg followed by intubation using a reinforced endotracheal tube. Invasive monitors were a must; arterial line and central venous line were inserted as well as transesophageal echocardiography. The patient was in prone position, and the procedure lasted 6 h. PRBCs 15 ml/kg were transfused during the procedure, anesthesia was maintained by TIVA using propofol and remifentanil. The procedure was uneventful otherwise. At the end, the endotracheal tube was replaced by a regular one and the child was shifted to the Pediatric Intensive Care Unit (PICU) intubated. The patient was extubated next day, after 3 days, she was transferred to the ward and after 17 days, she was discharged home.

Seven months later, at the age of 5 years and 3 months, the same patient had growing rods expansion, a procedure which lasted 1 h. She had smooth IV induction using propofol 2 mg/kg, fentanyl 1.5 mcg/kg, and rocuronium 0.6 mg/kg then intubated. The patient was extubated at the end of the procedure and shifted to the PICU. She was transferred to the ward next day and went home after 3 days.

Seven months later, at the age of 5 years and 10 months, the patient had another growing rod expansion which was also uneventful. She was extubated and transferred to HDU. The total length of the hospital stay was 6 days.

At the age of 6 years and 7 months, this patient had dental rehabilitation under GA which was uneventful, she was extubated, sent to the ward for 1 day and then discharged home.

Again, at the age of 7 years and 7 months, another growing rod expansion was done. She had a smooth IV induction using fentanyl and propofol. There were no invasive monitors. After an uneventful procedure, the patient was extubated, sent to the ward, and after 3 days, she was discharged home.

**Case 2**
The second patient was a male who is known to have Noonan’s syndrome and hypoplastic left heart syndrome. He had the Norwood procedure done in the UK; later he underwent Glenn procedure then a fenestrated Fontan. During early periods after surgery, he had cyanosis on exertion, bilateral femoral, and right internal jugular vein thrombosis. Furthermore, he had long-term ICU stay and developed subglottic stenosis. Echocardiography showed mild depression of left ventricular (LV) function, patent arch, fenestration, Glenn, and Fontan.
The first procedure was dental rehabilitation before device closure of Fontan fenestration. This was 2 years after Fontan procedure, he was 5 years old, his weight was 15.5 kg, and he was on aspirin and lisinopril. He was connected to the ASA standard monitors, received antibiotic prophylaxis against endocarditis; intubation was done after smooth IV induction using propofol and fentanyl. The procedure was uneventful. At the end, he was transferred to the ward where he was admitted for 1 day then discharged home.

The next dental rehabilitation was done 5 years later at the age of 10 years. The procedure was done as a day case. He had an IV induction. The procedure was uneventful. At the end of the procedure, the patient was discharged home.

**Case 3**

The third patient is an 8-month-old male, his primary CHD was pulmonary atresia (PA) with intact ventricular septum, RV hypoplasia. He underwent Glenn procedure at 4 months of age and was posted for elective circumcision as a 1-day procedure. The preoperative evaluation showed good functional capacity and LV function. After applying ASA monitoring standard, inhalation induction was done; laryngeal mask airway was inserted to secure the airway followed by caudal analgesia. The procedure was uneventful, and he was discharged home on the same day.

**Case 4**

The fourth patient is an 11-year-old female, with the Fontan circulation for the past 3 years and 10 months. Her primary CHD was double inlet LV with PA. She had mild aortic regurgitation and was posted for elective adenotonsillectomy. She had reasonable functional capacity and ventricular function preoperatively. Intraoperatively, there were no significant issues documented; Standard ASA monitors were used with no invasive ones, she had intubation after IV induction and was extubated at the end of the surgery. She was admitted to the ward for 1 day after the procedure.

**Discussion**

Single ventricle patients survive nowadays to older ages thank to the improvement in surgical and palliative procedures.

It is not uncommon to face such patients presenting for various procedures under sedation or general anesthesia, and consequently, it is not always feasible to provide their anesthesia services in specialized cardiac centers by pediatric cardiac anesthesiologists.

It is equally important for the anesthesiologist, surgeon, and all who are involved in perioperative care for these patients to know their nature, pathophysiology, and management keeping patient safety as a priority under all circumstances.

This knowledge will avoid unnecessary cancellation, rescheduling, and unnecessary admissions to ICU. Hence, the aim of presenting our case series is to highlight significant points and tips that need more exploration and study.

We report here ten procedures done for four patients who have SVP at different palliative stages. Out of ten operations, there were six of them done for one patient.

There was a variation for the same patient at the same stage of palliation may be for the same procedure; in management, the use of invasive monitors and disposition postoperatively whether intubated or extubated, to PICU or the ward or directly home.

Analyzing our results and variability in levels of monitoring intraoperative and disposition postoperative, we can point at several factors that may be involved and may be contributing and they are under one of three arms; first is the surgical procedure, its complexity, invasiveness, duration, and extent of fluid shift and blood loss is a factor.

The second is the patient, his general condition, age, weight, his cardiac problem, stage of palliation, and functional status.

The third is the operators, more importantly, the expertise of anesthetist and background which may vary from overdoing to underestimation of the situation.

Faraoni et al. recently developed a risk stratification score for children with CHD undergoing noncardiac surgery, and they identified those with SVP to be at high risk for perioperative complications, they revealed eight predictors for in-hospital mortality.[4]

Ramamoorthy et al. collected data from the pediatric perioperative cardiac arrest (CA) registry[5] and reported that the most common category of HD lesion in patients suffering CA was single ventricle 19%. In the same study, they noted that three-quarters of the CA in patients with HD occurred in children younger than 2 years old.[6]

In a retrospective study, Brown et al. revealed an increased incidence of adverse advents in patients with SVP undergoing noncardiac procedures.[7] They recommended that these patients should be managed by an experienced and dedicated team of pediatric anesthesiologists at a tertiary medical center where cardiac surgeons, invasive cardiologists, and
pediatric intensivists are readily available to manage and prevent critical events promptly.[8]

On the other hand, White and Peyton, in their review classified children with CHD for noncardiac procedures into high-risk, intermediate-risk, and low-risk groups [Table 1].[8] At the end, they recommended that high-risk children require transfer to a specialist center because full pediatric intensive care and cardiology services may be needed. Depending on the locally available facilities and expertise, intermediate-risk children should be discussed with the specialist center and should consider the transfer.[8]

Low-risk patients may be operated in the local hospital, and preoperative assessment, induction, and maintenance should all be individualized to the child and tailored to the type of surgery.[8]

Watkins et al. identified independent risk factors for the clinical outcomes of noncardiac operations in children with complex CHD. Patients in the stage before Glenn, going more invasive procedures or receiving inotropes, angiotensin-converting enzyme inhibitors, or digoxin appear to be at risk.[9]

Patients who have been hospitalized for more than 14 days appear to be at higher risk for requiring postoperative mechanical ventilation and with ventricular dysfunction, receiving inotropes, and not receiving digoxin may be at risk for protracted hospitalization.[9]

Despite the increased risk for these children, there is no clear method to identify, stratify, or adjust and improve that risk in the preoperative period.

Here, in our series, we drew attention that more studies are required to identify risk factors and find a system that integrates these risk factors and produce a severity scoring system that identifies patients with higher risk and allow for better preparation and improved outcome.

We have started working in our institution on a project aiming to come out with predictors of ICU admission in such patients and find a strategy that may help to improve outcome.

**Declaration of patient consent**
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and anonymity cannot be guaranteed.

**Financial support and sponsorship**
Nil.

**Conflicts of interest**
There are no conflicts of interest.

**References**

1. Perloff JK, Warnes CA. Challenges posed by adults with repaired congenital heart disease. Circulation 2001;103:2637-43.
2. Yuki K, Casta A, Uezono S. Anesthetic management of noncardiac surgery for patients with single ventricle physiology. J Anesth 2011;25:247-56.
3. Hennein HA, Mendeloff EN, Cilley RE, Bove EL, Coran AG. Predictors of postoperative outcome after general surgical procedures in patients with congenital heart disease. J Pediatr Surg 1994;29:866-70.
4. Farasone D, Yeo D, Nasr VG, Diniardo IA. Development and validation of a risk stratification score for children with congenital heart disease undergoing noncardiac surgery. Anesth Analg 2016;123:824-30.
5. Murray JP, Geiduschek JM, Ramamoorthy C, Haberkern CM, Hackel A, Caplan RA, et al. Anesthesia-related cardiac arrest in children: Initial findings of the pediatric perioperative cardiac arrest (POCA) registry. Anesthesiology 2000;93:6-14.

---

**Table 1: Risk classification of children with heart disease undergoing noncardiac surgery**

| High Risk                                                                 | Intermediate Risk                                                                 | Low Risk                                                                 |
|--------------------------------------------------------------------------|---------------------------------------------------------------------------------|-------------------------------------------------------------------------|
| Physiologically poorly compensated and/or presence of major complications | Physiologically normal or well compensated                                        | Physiologically normal or well compensated                              |
| a. Cardiac failure                                                         |                                                                                 |                                                                         |
| b. Pulmonary hypertension                                                 |                                                                                 |                                                                         |
| c. Arrhythmias                                                            |                                                                                 |                                                                         |
| d. Cyanosis                                                               |                                                                                 |                                                                         |
| Complex lesions (single-ventricle or balanced circulation physiology, cardiomyopathy, aortic stenosis) | Simple lesions                                                                  | Simple lesions                                                          |
| Major surgery (intraperitoneal, intrathoracic, anticipated major blood loss requiring transfusion) | Major surgery (intraperitoneal, intrathoracic, anticipated major blood loss requiring transfusion) | Minor surgery                                                          |
| Under 2 yr old                                                            | Under 2 yr old                                                                   | Over 2 yr old                                                          |
| Emergency surgery                                                         | Emergency surgery                                                                | Elective Surgery                                                       |
| Preoperative hospital stay more than 10 days                              | Preoperative hospital stay more than 10 days                                     | Preoperative hospital stay < 10 days                                    |
| ASA physical status IV or V                                                | ASA physical status IV or V                                                      | ASA physical status I or III                                           |

---

---
6. Ramamoorthy C, Haberkern CM, Bhananker SM, Domino KB, Posner KL, Campos JS, et al. Anesthesia-related cardiac arrest in children with heart disease: Data from the pediatric perioperative cardiac arrest (POCA) registry. Anesth Analg 2010;110:1376-82.

7. Brown ML, DiNardo JA, Odegard KC. Patients with single ventricle physiology undergoing noncardiac surgery are at high risk for adverse events. Paediatr Anaesth 2015;25:846-51.

8. White MC, Peyton JM. Anaesthetic management of children with congenital heart disease for non-cardiac surgery. Contin Educ Anaesth Crit Care Pain J 2012;1:12.

9. Watkins SC, McNew BS, Donahue BS. Risks of noncardiac operations and other procedures in children with complex congenital heart disease could assist clinicians in assessing perioperative risk. Ann Thorac Surg 2013;95:204-11.