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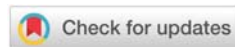
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### Review Article

## Risk assessment and anesthesia management in children with congenital heart disease undergoing non-cardiac surgery

### Abstract

The prevalence of congenital heart disease is about 8 to 10 case per 1000 live births and is a major cause of increased mortality and morbidity in pediatric patients undergoing noncardiac surgery. Therefore safe anesthesia and adequate recovery should be provided. It is important to determine the patient's risk score in the preoperative period. However, the risk assessment tools have a limited prediction for increased mortality and morbidity of non-cardiac surgery. The most important point in determining the anesthesia method is to be aware of the latest situation both anatomically and physically about the circulation of patient and to create the specific planning. In these patients, the aim of maintenance of anesthesia is to increase arterial oxygen saturation by increasing pulmonary blood flow. Thus the use of appropriate anesthesia and monitoring methods through multidisciplinary decision-making and planning, as well as the identification of high-risk patients based on risk classification, may reduce mortality and morbidity in the pediatric patients with congenital heart disease.

### Introduction

Congenital heart disease is a cause of increased mortality and morbidity in pediatric patients undergoing noncardiac surgery. In United States the prevalence of congenital heart disease is about 8 to 10 case per 1000 live births [1]. Hemodynamic instability in the perioperative period, increased need for postoperative mechanical ventilation and prolonged hospital stay are the most common complications in these patients. Proper preparation is necessary before surgery in order to reduce the incidence of such complications. The main objectives are to perform procedures in a fully-equipped medical center, with proper consultations and appropriate monitoring. Safe anesthesia and adequate recovery should be provided by an experienced anesthesia team. Today, 90% of children with congenital heart disease are able to reach the adult period thanks to prenatal diagnosis and interventions, improvements in surgical techniques, and improved intensive care support [2]. 30% of these patients undergo noncardiac surgery at least once until 5 years of age due to concomitant trachea-esophageal fistula, anorectal anomalies, cleft palate, lip or non-cardiac anomalies originating from the renal system. Several interventions in these patients may lead to undesirable hemodynamic responses.

In the National Surgical Quality Improvement Project (Pediatrics) study conducted between 2013 and 2015, the pneumoperitoneum model used in laparoscopic surgeries compared the open surgical method with the 30-day mortality, morbidity and postoperative hospital stay, and the patients undergoing laparoscopic surgery have demonstrated superiority in terms of 30-day mortality, hospital mortality, hospital stay, and decreased blood transfusion in patients with congenital heart disease [3]. In a study conducted in a group of patients undergoing non-cardiac surgery, 22% of patients with congenital heart disease developed cardiac arrest, and half of them occurred during non-cardiac procedures [4]. The mean age of these patients was below 2 years of age and half of the patients were younger than 6 months. 24% of patients with cardiac arrest were identified as single ventricle, 26% as outflow obstruction and 18% had left to right shunt. The presence of ventricular dysfunction with the use of preoperative angiotensin converting enzyme (ACE) inhibitor, inotropic agents and digoxin were found to be associated with prolonged hospital stay in the postoperative period. On the other hand, several airway abnormalities may complicate the course of congenital heart disease [5]. Besides, during resuscitation, pediatric tracheal intubation requires experience and training, thus presents a high incidence of complications [6].

Systemic vascular resistance changes caused by general anesthetic agents reduce pulmonary blood flow in the presence of shunt. The idea that the blood pressure described by the Ohm's law is proportional to the heart rate and stroke volume may not be valid in these patients. Because it will not increase the discharge volume of the ventricle with a limited capacity of fluid loading to increase the stroke volume.

### Risk assessment

The American Society of Anesthesiologist's Physical Status Score (ASA-PS) and NARCO-SS (neurological, airway, respiratory, cardiovascular, surgical score) are used in the pediatric patient group to determine the patient's risk score in the preoperative period [7]. However, as in pediatric patients with congenital heart disease, they have a limited prediction for increased mortality and morbidity of non-cardiac surgery. Pediatric Perioperative Cardiac Arrest (POCA) study found that perioperative cardiac arrest was present in 34% of patients with congenital heart disease undergoing noncardiac surgery in a 11-year period. The most important factors that determine the risk for the surgical procedure of these patients were determined as the patient's age, severity of cardiac anomaly and accompanying comorbidities. In another retrospective analysis, 47% of the patients were found to have a prolonged intensive care stay after elective noncardiac surgery [8]. However, the variety of heart disease and surgical procedures leads to difficulties in determining the patient's risk. In different studies, the complexity of heart disease, type of cardiac surgery and the functional capacity of the patient have been shown to be effective on postoperative complications. In order to determine the appropriate anesthesia approach for both pathophysiological and anatomical differences, risk assessment should be performed to reduce the complications and to ensure the efficient use of resources [9]. For this purpose, the pediatric patients with congenital heart disease were evaluated in the Risk Adjustment for Congenital Heart Surgery Score (RACHS-1), Aristotle Basic Complexity Score (ABC score) and Society of Thoracic Surgeons and the European Association for Cardiothoracic Surgery Mortality (STS-EACTS) scoring systems are used and are more specific to cardiac surgery. Several patients have associated diseases such as esophageal atresia. Waterson risk classification is used for the perioperative risk assessment [10].

Faraoni et al. [11], developed a risk model based on patient data from the records of the American College of Surgeons National Surgical Quality Improvement Program which included 4375 pediatric patients between 2012-2014. In addition to inotropic agent support, mechanical ventilation, cardiopulmonary resuscitation and chronic kidney damage, which are considered as preoperative critical disease markers; the severity of heart disease and the type of lesion were determined as an independent predictor of in-hospital mortality following non-cardiac surgery. In another study, patients were studied in three groups: high, medium and low risk [9].

### Preoperative period

American Heart Association 2007 recommendations for

the prophylaxis of infective endocarditis in these patients are currently being applied and are recommended only for patients at high risk. These include: a previous attack of endocarditis, heart valve repair (mechanical or biological prosthesis), presence of congenital heart disease, uncorrected cyanotic congenital heart disease, corrective or palliative prosthetic material or catheterization, congenital heart disease with corrected but residual anomaly, and heart transplantation candidates [12]. In these patients, the coagulation profile, Hb / Htc ratio and electrolyte status are essential. The increased Hb / Htc ratio is in favor of chronic hypoxia. An Hb level of > 20 g/dL or Htc > 65% may lead to hyperviscosity and decreased capillary blood flow and perfusion, leading to reduced oxygen delivery to the perioperative tissues and thromboembolic complications such as stroke. The target Htc level should be a maximum of 60%. The effect of chronic liver congestion on the production of coagulation factors in patients with Fontan circulation should also be noted. Unfortunately, phlebotomy does not reduce the risk of stroke in these patients. Prolonged preoperative fasting period also contributes to hypoviscosity and coagulopathy due to secondary polycythemia.

Premedication is necessary in this group of patients whose hemodynamics can rapidly deteriorate, but the increase in PaCO<sub>2</sub> with respiratory depression that may develop in deep sedation may increase pulmonary artery resistance and cause hypoxemia to be aggravated. Therefore, it is recommended to administer 0.5 mg/kg of oral midazolam preoperatively 15-30 min before the procedure.

Congenital heart disease can be *accompanied* with difficult airway management [13]. The Italian Difficult Airways Study Group, on behalf of the Italian Society for Anesthesia and Intensive Care (SIAARTI), has recommendations on pediatric difficult airway as a separate pediatric section [14].

### Monitorization

Due to the negative inotropic effects of anesthetic agents, hemodynamic monitoring is mandatory except for minor surgery and interventions. In patients with a Blalock-Taussig shunt, blood pressure measurements on the same side will yield lower results, so measurements from the opposite side arm should be performed. Temperature monitoring is mandatory for avoiding hypothermia, maintenance of peripheral perfusion in polycythemic patients and prevention of metabolic acidosis. The use of ultrasonography in central venous catheterization allows the reduction of possible thromboembolic complications.

### Anesthesia management

The most important point in determining the anesthesia method is to be aware of the latest situation both anatomically and physically about the circulation of patient and to create the specific planning. In these patients, the aim of maintenance of anesthesia is to increase arterial oxygen saturation by increasing pulmonary blood flow. No anesthetic agent is contraindicated in this patient group. In the presence of a right-to-left shunt, there is an increase in minimum alveolar concentration and a prolonged inhalation induction due to decreased pulmonary

blood flow. In contrast, the induction with intravenous agents is rapid. Although bradycardia is common, routine prophylaxis prior to induction is not recommended.

Hemodynamic stabilization can be achieved with a single dose of Fentanyl (25–30 mcg/kg) in the presence of inadequate ventricular function for induction of anesthesia. Hypnotic agents (Propofol 1–3 mg/kg) and Midazolam 0.1 mg/kg can be used in patients with intact ventricular function. The choice of muscle relaxant agent is recommended according to the duration of operation. Rocuronium, vecuronium, cisatracurium and mivacurium are frequently used. Rocuronium seems to be advantageous because it can be neutralized with Sugammadex. In these patients, neuraxial blocks can be applied, as long as the sudden decrease in peripheral vascular resistance can be prevented.

Pulmonary blood flow in patients with Fontan and Glen circulation; the pressure gradient is determined by the relationship between central venous pressure and pulmonary vascular resistance. As a result of myocardial pump function deterioration, atrial pressure increases and SpO<sub>2</sub> decreases with decreasing pressure gradient in pulmonary circulation. SpO<sub>2</sub> and cardiac output are further reduced by the blood flow to the lungs, resulting in a vicious cycle.

The application of PEEP results in an increase in intrathoracic pressure with a reduction in the preload and cardiac output, and is significant in patients with Fontan circulation. The pneumoperitoneum model, which leads to an increase in intraabdominal pressure, can reduce cardiac output by reducing the venous return. In this case, adequate volume delivery and appropriate ventilation support play a key role in maintaining haemodynamic stability.

Symptoms such as Fontan's circulation, severe pulmonary hypertension, cyanosis and heart failure, valvulopathy, chronic anticoagulation therapy and arrhythmia are evaluated in the high-risk group. The American Heart Association (AHA) and the American College of Cardiology (ACC) reported that patients without these symptoms do not require a specialized center for non-cardiac interventions [15].

## Conclusions

The use of appropriate anesthesia and monitoring methods through multidisciplinary decision-making and planning, as well as the identification of high-risk patients based on risk classification, may reduce mortality and morbidity in the pediatric patients with congenital heart disease.

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