UDK: 616-089.5-053.2:616.12-039

ANESTHESIA FOR NON-CARDIAC SURGERY IN CHILDREN WITH CONGENITAL HEART DEFECTS

Malinovska-Nikolovska L[1], Chadikovski V[1], Krmzova-Gjorgioska M[1]

[1]*Department of Pediatric Cardiac Surgery, "AcibademSistina" Clinical Hospital, Skopje*

Abstract

Congenital heart defects are the most common form of birth defects and occur in over 1% of newborns. Patients with congenital heart disease undergoing noncardiac surgery have increased perioperative morbidity and mortality. The purpose of this lecture is to teach anesthesiologists, who do not usually deal with cardiac anesthesia in children with congenital heart defects, to provide reliable and safe perioperative care for these patients. The anesthesiologist is required to have basic and advanced knowledge of the heart defect and its pathophysiological characteristics. In the preoperative preparation, an exhaustive anamnesis and status is required, as well as contact of the anesthesiologist with the child's primary pediatric cardiologist. Prevention of endocarditis should always be considered. The administration of anesthesia is in accordance with the pathophysiological characteristics of the congenital heart defect, and the underlying surgical disease for which the child is operated. Certain patients need to be sent to a specialized center, where, in addition to basic surgery, they also have teams to care for these patients. The anesthesiologist needs to know the anatomy and physiology of the congenital heart defect, and correctly to assess the current condition of the patient, in order to provide safe perioperative care in these patients.

Key Words: *anesthesia, congenital heart defects, non-cardiac surgery.*

Introduction

Congenital heart defects (CHD) are the most common form of heart defects and occur in over 1% of newborns. The prognosis, especially in patients with complicated CHD, has improved significantly in recent decades. Not only diagnostics, but also treatment, such as surgery, interventional procedures and intensive care medicine, have made significant progress in the last few decades. That's why it is expected that an increasing number of patients with palliative or corrective operations of CHD will need non-cardiac surgery.

Regarding the risks, there are studies that confirm that the risk of perioperative cardiac arrest is higher in children with CHD, but on the other hand, the frequency of perioperative complications in non-cardiac surgery in patients with CHD who do not have pulmonary hypertension, congestive heart failure or cyanosis are low, same as in patients who do not have CHD. The purpose of this lecture is that anesthesiologists who do not usually deal with cardiac anesthesia in

CHD, can safely and reliably manage the perioperative care of patients with CHD, having a basic and advanced knowledge of CHD and its pathophysiological characteristics.

Special Aspects of the Medical Examination and Anamnesis in Patients with VSM

The anesthesiologist, who examines the patient with CHD and informs the parents and the child about the perioperative anesthetic care, must have at least a basic knowledge of CHD and its pathophysiological characteristics. Parents are usually very well informed about their children's CHD and know their medical history well, including therapy and surgeries that the patients have undergone. That is why an interview with the parents gives us a lot of information. But in addition, of course, the last examination by a cardiologist or cardiac surgeon must be checked, in order to understand the real status of the patient and the anatomy of their circulatory system.

In 30% of children with CHD there are also other extracardiac anomalies, and this is significantly more than the prevalence in children without CHD. The presence of these extracardiac malformations significantly increases the perioperative risk. Therefore, a detailed history and examination, especially in younger patients, is particularly important. In addition to this, it is necessary to assess the degree of heart failure. The questions are asked depending on the age of the child: Does the baby sweat while drinking from a bottle? Is it gaining weight? Does it have respiratory infectionsfrequently? Does the child tire easily when playing? In this way, the content of the questions changes according to the age of the child. The NYHA (New York Heart Association) functional classification is also useful in assessing the patient's abilities (Table1).

Table 1. Functional classification New York Heart Association (NYHA).

NYHA Class	**Symptoms**
I	There is no restriction on physical activity. Ordinary physical activity does not cause excessive fatigue, palpitations, dyspnea.
II	Mild restriction of physical activity. Comfortable at rest. Ordinary physical activity results in fatigue, palpitations, or dyspnea.
III	Significant limitation of physical activity. Comfortable at rest. Little activity causes fatigue, palpitations, or dyspnea.
IV	Any physical activity causes discomfort. Symptoms at rest. If any physical activity is undertaken, the discomfort increases.

It is also very important to assess the possibility of a difficult airway. Laryngotracheal stenosis can cause a difficult airway. It is important to ask how long mechanical ventilation lasted after heart surgery and whether there are symptoms of possible stenosis. Certain syndromes are also associated with both difficult airway and CHD.

Prevention of Endocarditis

Infective endocarditis is a life-threatening disease and is difficult to treat, so it can have serious consequences for patients. For this reason, liberal antibiotic treatment has been recommended and implemented for all patients with CHD for a long time. However, it is now recommended that only defined patients at high risk of endocarditis receive antibiotic treatment. Table 2 shows patients at risk for endocarditis who should receive prophylaxis, and Table 3 provides an example of antibiotic prophylaxis.

Table 2. Patients at risk for endocarditis.

Heart valve operations (mechanical or biological valve)	
Previous endocarditis	
Congenital heart defect (CHD)	Uncorrected cyanotic CHD
	During the first 6 months after palliative or corrective surgeries using prosthetic material or catheter intervention
	Corrected VSM with residual damage to the prosthetic material
Heart transplant patients who develop cardiac valvulopathy	

Table 3. Example of antibiotic prophylaxis for endocarditis for non-cardiac surgery in high-risk patients.

Method of application	**Antibiotic**	**Dose**	
		Young child	**Older child**
Oral (PO)	Amoxicillin	50mg/kg PO	2gr PO
Intravenous IV (when oral not possible)	Cephazolin	50mg/kg IV	2gr IV
In penicillin allergies, oral PO	Clindamycin	20mg/kg PO	600mg PO
In penicillin allergies,IV	Clindamycin	20mg/kg IV	600mg IV

Laboratory Tests

Patients with cyanotic CHD have very high Hb/Hct as a consequence of chronic hypoxia. Hb>20gr/dL or Hct>65% leads to a high risk for thromboembolic complications. Hyperviscosity can be aggravated by too long preoperative fasting or vomiting, or insufficient intraoperative fluid replacement. For this reason, the patients with cyanotic CHD should receive adequate fluid replacement.Also, during intraoperative bleeding, the target hematocrit is always higher than in other children (about 40-45%), because the chronic hypoxia must be compensated in this way.Chronic congestion of the liver can reduce the production of coagulation factors, for example in patients with Fontan circulation.Preoperatively, serum electrolytes should be checked in patients receiving diuretics. The EKG will show us the rhythm of the patient and the workload of the heart. A chest X-ray shows the position of the heart, size, possible atelectasis, acute respiratory infections, and the position of the hemidiaphragms.

Premedication in Patients with CHD for Non-cardiac Surgery

Premedication is especially important for patients with CHD, because hemodynamic balance can be disturbed due to stress, fear due to separation from parents, or induction of anesthesia. Some patients with CHD have had multiple heart surgeries and therefore some of them have unpleasant memories and impressions. Premedication is a solution to this problem. However, excessive sedation must be avoided, which may result in an increase in $PaCO_2$ due to respiratory suppression with subsequent hypoxia. This is especially important in patients with pulmonary hypertension.

Oral application of Midazolam 0.5mg/kgTT 15-30 minutes before entering the operating room is commonly used for premedication. Oral premedication with midazolam is also safe and effective in children with cyanotic CHD. If we have set up an intravenous route, intravenous application of Midazolam 0.05-0.1mg/kgTT and Ketamine 1mg/kgTT can be used, but still after giving the drugs i.v., monitoring of the child is required and the possibility of possible oxygen therapy if it is necessary. Ketamine can be given both orally and intramuscularly. Dexmedetomidine intranasal 1mcg/kgTT 45-60minutes before intervention can also be used.

Intraoperative Management of Patients with CHD

All used drugs and methods of induction of anesthesia can be used in patients with CHD, but of course in a certain dose and combination. We should always think about systemic vascular resistance (SVR) and pulmonary vascular resistance (PVR). Inhalation anesthesia and inhalation induction of anesthesia can be used in these patients, but caution should be exercised with higher doses in patients with poor cardiac reserve. All intravenous anesthetics can be used, but be careful with propofol, which strongly reduces SVR. Ketamine is the drug of choice in patients with weak cardiac reserve, but it must not be used in high doses, because it has a negative inotropic effect. High-dose opioid anesthesia is suitable for these patients, but the problem of postoperative respiratory depression and patient extubation remains.

The amount of intraoperative monitoring will depend on the severity of the CHD, but also on the degree of risk of the operation itself. Routine monitoring consisting of ECG, noninvasive arterial

pressure, SpO2, and temperature is sufficient for the most interventions. But if we have high-risk operations and/or complex heart defects, it is necessary to place a central venous catheter, an arterial catheter, as well as measurement of diuresis and NIRS.

Congenital heart defects with left-right shunt are present in 50% of these children. Examples are atrial septal defect, ventricular septal defect, A-V channel. Here the blood from the left heart, instead of going completely through the aorta in the systemic circulation, partially, through the existing communication, returns to the right heart and burdens the pulmonary circulation. Use of 100% O2 should be avoided in these patients, as it is a potent vasodilator of the pulmonary vessels and may lead to pulmonary congestion.

The existence of a right-left shunt means that the blood from the right side of the heart passes partially into the left heart through the existence of communication. We have this in the Tetralogy of Fallot. After induction of anesthesia, as a result of a drop in SVR due to the existence of a VSD, the normally left-to-right shunt now becomes right-to-left, as the pressure in the right ventricle becomes higher than the pressure in the left ventricle. The patient becomes cyanotic after induction of anesthesia. This is treated by giving a fluid bolus and increasing SVR with phenylephrine and reducing infundibular spasm with B blockers. Another important aspect in these patients is that pulseoximetry is higher than real, as saturation decreases, and end-tidal CO2 is different from arterial CO2. Therefore, taking gas analyzes in these patients is mandatory.

A mean pulmonary arterial pressure greater than 25mmHg at rest and 35mmHg during activity is pulmonary hypertension. The high pulmonary flow that occurs with an unrestricted left-to-right shunt will lead to congestive heart failure and pulmonary hypertension. Initially, pulmonary hypertension is reactive and responds to hypothermia, stress, pain, acidosis and hypercarbia, hypoxia and increased intrathoracic pressure, but later it becomes fixed. Prevention of these trigger factors should be the goal of anesthetic management. A crisis of pulmonary hypertension can occur in the case of shallow anesthesia and airway instrumentation. Its termination requires deepening of anesthesia, administration of 100% O2, good CO2 elimination, and ventricular support with inotropic support.

Patients with single-chamber pathology go through several stages of palliative operations. B-T shunt is performed in newborn age, then Glenn operation and finally Fontan operation. Children with B-T shunt and Glenn have a normal saturation of about 70-85%. In patients with Fontan surgery, the venous inflow into the heart is directed directly into the pulmonary artery, and the entire heart, as a single chamber, works only to pump blood through the aorta into the systemic circulation. Venous blood flow will depend on the gradient between central venous pressure and pulmonary vascular resistance. Therefore, these patients need easy lung ventilation, without the use of high PEEP and high respiratory pressures, as well as improvement of the preload with a sufficient amount of fluids. These patients are volume dependent. Mechanical ventilation negatively affects these patients, so it is necessary to plan their early extubation.

Risks Classification

Children with BSM who have noncardiac surgery are at increased risk for morbidity and mortality. The most important factors affecting the risk are the complexity of the congenital heart

defect, the physiological status of the child, the type of surgery and the age of the child. To enable a practical and structured approach to dealing with these patients, the risk for surgery is classified into three groups, shown in Table 4.

Table 4. Risk classification of children with congenital heart defects for non-cardiac surgery.

High risk	**Medium risk**	**Low risk**
Physiologically poorly compensated or presence of major complications	Physiologically normal or well compensated	Physiologically normal or well compensated
a) Heart failure		
b) Pulmonary hypertension		
c) Arrhythmias		
d) Cyanosis		
Complex lesions (univentricular heart, cardiomyopathy, aortic stenosis)	Simple lesions	Simple lesions
Major surgery (intraperitoneal, intrathoracic, anticipated major blood loss)	Major surgery (intraperitoneal, intrathoracic, anticipated major blood loss)	Minor surgery
Under 2 years of age	Under 2 years of age	Over 2 years of age
Emergency surgery	Emergency surgery	Elective surgery
Preoperative hospital stay more than 10 days	Preoperative hospital stay more than 10 days	Preoperative hospital stay less than 10 days
ASA status IV or V	ASA status IV or V	ASA status I-III

Conclusion

Children with congenital heart defects presenting for noncardiac surgery are at increased risk for perioperative morbidity. High-risk children require transfer to a specialized center, where there is specialized cardiosurgical intensive care and cardiology. Children with intermediate risk, depending on the circumstances, can be operatedatthe local center, but with specialist support from a specialized center or possible transfer. Low-risk children can be operated at the local hospital. All anesthesiologists responsible for children with congenital heart defects need to understand the anatomy, physiology and risk factors associated with the perioperative morbidity, perform a good preoperative assessment, and know the impact of anesthetics and mechanical ventilation in this patients' population.

References:

1. 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. AnesthAnalg2010;110:1376-82.
2. Nasr VG, Staffa SJ, Faraoni D, DiNardo JA. Trends in mortality rate in patients with congenital heart disease undergoing noncardiac surgical procedures at children's hospitals. Sci Rep. 2021;11:1543.
3. Taylor D, Habre W. Risk associated with anesthesia for noncardiac surgery in children with congenital heart disease. PaediatrAnaesth. 2019;29:426–434.
4. Faraoni D, Vo D, Nasr VG, DiNardo JA. Development and validation of a risk stratification score for children with congenital heart disease undergoing noncardiac surgery. AnesthAnalg. 2016;123:824–830.
5. Nasr VG, DiNardo JA, Faraoni D. Development of a pediatric risk assessment score to predict perioperative mortality in children undergoing noncardiac surgery. AnesthAnalg. 2017;124:1514–1519.
6. White MC, Peyton JM. Anaesthetic management of children with congenital heart disease for non-cardiac surgery. ContinEducAnaesthCrit Care Pain 2012;12:17-22.
7. Menghraj SJ. Anaesthetic considerations in children with congenital heart disease undergoing non-cardiac surgery. Indian J Anaesth 2012;56:491-5.
8. Thiene G, Frescura C. Anatomical and pathophysiological classification of congenital heart disease. CardiovascPathol 2010;19:259-74.
9. Brown ML, DiNardo JA, Odegard KC. Patients with single ventricle physiology undergoing noncardiac surgery are at high risk for adverse events. PaediatrAnaesth 2015;25:846-51.