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General principles of anesthesia in pediatric cardiac surgery

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Abstract

Cardiac anomalies, defined as structural abnormalities of the heart or intrathoracic vessels, taking all the different anatomic forms together, are some of the most common congenital anomalies at birth. It is estimated that between two and 10 in every 1,000 LB are affected by some type of cardiac malformation. One third of those affected have critical cardiac malformations, defined as those that require immediate investigation and treatment, or those that will lead to death during the first year of life. As medicine has advanced and early detection become possible, many of these newborn will survive and contribute to increase the population of adults with congenital heart disease. Currently, there are approximately more than 600 children and adults with congenital heart disease who present for cardiac surgery annually in prince sultan cardiac centre. Those patients have multiple congenital variations leading to an incremental risk for surgical procedures. The aim of this review is to discuss current trends concerning anesthesia in children with congenital heart disease in our center and to share with the readers our clinical profile of anesthesia in pediatric cardiac surgery.

INTRODUCTION

Congenital heart diseases are classified into:

CYANOTIC HEART DISEASE

Cyanotic heart disease is a term for a group of defects in the structure or function of the heart or great vessels, preventing normal blood flow from the right part to the left part of the circulatory system. This abnormal blood flow results in cyanosis, poor oxygenation of the body that causes a bluish coloration in the lips, fingers, and toes during exercise. Types of cyanotic heart diseases are:

1. Tetralogy of Fallot
2. Transposition of the great arteries (vessels) (TGA)
3. Ebstein anomaly
4. Tricuspid atresia (a deformity of the tricuspid heart valve)
5. Total anomalous pulmonary venous return (TAPVR)
6. Pulmonic stenosis
7. Truncus arteriosus
8. Hypoplastic left heart syndrome (HLHS)

ACYANOTIC HEART DISEASE

Acyanotic heart disease is a broad term for any congenital heart defect in which there is a defect in one of the walls separating the chambers of the heart, or an obstruction to one valve or artery. This problem occurs just before or after birth and may cause problems for newborns. Types of acyanotic heart disease are:

1. Patent ductus arteriosus (PDA)
2. Coarctation of the aorta
3. Atrial septal defect (ASD)
4. Ventricular septal defect (VSD)
5. Atrioventricular septal defect (AVSD)
6. Aortic stenosis (AS)
7. Pulmonary stenosis (PS)

CONGENITAL HEART DISEASES WITH LEFT-TO-RIGHT SHUNTS

Common left-to-right shunt lesions such as atrial septal defect (ASD), ventricular septal defect (VSD), patent ductus arteriosus (PDA), and endocardial cushion defect (ECD) and partial anomalous pulmonary venous return (PAPVR).

ASD occurs as an isolated anomaly in 5 to 10% of all congenital heart defects. It is more common in females. About 30% to 50% of children with congenital heart defects have ASD as part of the cardiac defect [1] . Spontaneous closure occurs more than 80% of the time in patients with defects between 3 and 8 mm before 1.5 years of age [1,2] . An ASD with a diameter >8 mm rarely closes spontaneously [2] .

VSD is about 2 per 1000 live births. VSD is the most common cardiac malformation. Spontaneous closure occurs in 30% to 40% of patients with membranous VSDs and muscular VSDs during the first 6 months of life [3,4] . These VSDs do not become bigger with age; rather, they decrease in size [5,6] .

PDA occurs in 5% to 10% of all congenital heart defects, excluding premature infants [7,8] . PDA in term infants results from a structural abnormality of the ductal smooth muscle rather than a decreased responsiveness of the premature ductus to oxygen [8] .

ECD occurs in 2% of all congenital heart defects. Of patients with complete ECD, 30% are children with Down syndrome. ECD is also a component of heart defects in asplenia or polysplenia syndrome [5] .

CONGENITAL HEART DISEASES WITH RIGHT-TO-LEFT SHUNTS

There are 6 common types of congenital heart diseases with right-to-left lesions: tetralogy of Fallot (TOF), transposition of the great arteries (TGA), truncus arteriosus (TA), tricuspid atresia, total anomalous pulmonary venous return (TAPVR), and pulmonary atresia with intact ventricular septum (PAIVS).

TOF is the most common intrinsic cyanotic congenital heart disease, which accounts for 5% to 10% of all congenital heart disease and has a prevalence of about 1 out of 2,000 live births [11] .

TGA is congenital heart lesion characterized by ventriculoarterial discordance. It accounts for 5% to 7% of all congenital cardiac malformations.

TA accounts for 1% to 2% of congenital heart disease [12] . There is a single semilunar valve and annulus with a common outflow trunk arising from the ventricles. It may be associated with DiGeorge syndrome [13] (Figure 8). Tricuspid Atresia accounts for 1% to 3% of congenital heart defects. It is congenital heart lesion characterized by agenesis of the tricuspid valve [14] .

TAPVR accounts for 1% of all congenital heart lesions. The pulmonary veins connect to the systemic venous system instead of to the left atrium.

PAIVS accounts for less than 1% of all congenital heart defects. The prevalence of this disorder is 4.1 per 100, 000 live births [15] .

In our institute >600 cardiac surgery procedures in adults and pediatrics are operated annually. Table 1 show the different pathology encountered.

Figure 1

Table 1: Pediatric cardiac anesthesia in prince sultan cardiac center for 2007(n=625).

No.	Surgery Type	Quantity	Percentage
1	Arterial Switch	10	1.6%
2	Aortic Aneurysmectomy release of aortic arch	1	0.16%
3	ASD Closure	28	4.48%
4	AV Canal Repair	28	4.48%
5	Bilateral PA Banding	1	0.16%
6	BT Shunt	45	7.2%
7	Central Shunt	4	0.64%
8	COA Repair	28	4.48%
9	Cortriatratium Repair	1	0.16%
10	DORV Repair	10	1.6%
11	Fontan	26	4.16%
12	Glenn	25	4%
13	Hemi-Truncus Repair	1	0.16%
14	IAA Repair	4	0.64%
15	LPA Reconstruction	1	0.16%
16	Mitral Valve Replacement	5	0.8%
17	Mitral Valve Repair	8	1.28%
18	Mustard Operation	3	0.48%
19	Norwood Operation	2	0.32%
20	PA Banding	58	9.28%
21	PA Reconstruction	5	0.8%
22	PAPVD Repair	2	0.32%
23	PDA Ligation	25	4%
24	Pulmonary Valve Replacement	23	3.68%
25	Rastelli Operation	1	0.16%
26	ROSS Operation	3	0.48%
27	Sub Aortic Membrane Resection	9	1.44%
28	TAPVD Repair	6	0.96%
29	TOF Repair	35	4.48%
30	Truncus Repair	2	0.32%
31	VSD Closure	59	9.28%

ANESTHESIA TECHNIQUE FOR PEDIATRIC CARDIAC SURGERY

The stress response to major cardiac surgery in infants and children is a subject that continues to evoke considerable interest. In their landmark study, published in 1992, Amand and Hickey [16] reported the use of sufentanil at a total mean dose of 37μ/kg, and an infusion that continued for 24 hours postoperatively, as a sole anesthetic for complex neonatal cardiac surgery. This technique was compared with halothane with morphine (mean dose of 0.35 mg/kg) intra-operatively, followed by intermittent intravenous doses of morphine and diazepam postoperatively. Stress response was evaluated by measuring changes in levels of adrenal hormones, cortisol, glucose, and lactate. This response was significantly decreased in the sufentanil group, and mortality and major complications such as sepsis and necrotizing enterocolitis were also significantly reduced. Duncan et al. [17] reported a dose response study of 2, 25, 50, 100, and 150µg/kg fentanyl before bypass in 40 children (average age 13 months). The patients who received 2µg/kg had significant increases in pre-bypass nor-epinephrine, glucose, and cortisol, and significantly higher heart rate and blood pressure than all other groups. Fentanyl doses of 25µg/kg or higher eliminated changes in all measured parameters for the duration of the surgery. Remifentanil is a synthetic, ultra-short-acting narcotic with a half-life of 3-5 minutes, independent of the duration of infusion, which is metabolized by nonspecific plasma esterases [18] . Donmez [19] reported a series of 55 children who underwent cardiac catheterization using a remifentanil infusion of 0.1µg/kg/min, with spontaneous respiration maintained. This technique provided cardiovascular stability, with minimal changes in heart rate, blood pressure or oxygen saturation. About half of the patients required additional sedation with midazolam. Apnea was infrequent, and time to recovery after discontinuation of remifentanil was only 2-4 minutes. Patients undergoing long cardiac catheterization procedures could thus potentially benefit from this agent. Its use has also been reported during surgical repair of atrial septal defects, with patients subsequently extubated in the operating room during switch operation the left ventricle is acutely converted from the pulmonary to the systemic ventricle. This imposes a significant increase in after load on the left ventricle and may predispose the patient to acute left ventricle dysfunction in the immediate post-operative period. Milrinone improves myocardial contractility, diastolic relaxation and decrease in after load due to vasodilatation. We conduct a study to determine the effect of milrinone infusion on hemodynamic profile in pediatric cardiac surgery. We found that milrinone infusion was shown to be more effective on a post-bypass course of hemodynamic profile by decreasing the LAP [40] .

ECHOCARDIOGRAPHY

Recently, echocardiographic techniques have been used to evaluate the ventricular function response to anesthetics in patients with congenital heart disease. A study comparing halothane, isoflurane, and sevoflurane [20] in 54 biventricular children, aged 1 month to 13 years and who underwent cardiac surgery, demonstrated that at both 1 and 1.5 minimum alveolar concentration {MAC) halothane, significant myocardial depression occurred. Sevoflurane preserved both CO and heart rate (HR), with fewer hypotensive and negative inotropic effects seen compared with halothane. The effects of these anesthetic agents on pulmonary (Qp) and systemic (Qs) blood flow in 30 biventricular patients with left-to-right shunts was also assessed echocardiographically [21] . Recently, the myocardial performance index (MPI) has been validated as an accurate assessment of ventricular function for all ventricular configurations, including functional single ventricle [22] . MPI is a useful new tool to assess ventricular function in response to anesthetics and other agents in this important and enlarging patient population [22]. In the near future three-dimensional echocardiography may also provide new insights into ventricular function in response to anesthetics in patients with congenital heart disease [23] . In a study by Ogletree and coworkers [24] , etomidate at therapeutic levels was shown to have negative inotropic effects in right ventricular muscle resected from patients with tetralogy of Fallot, and reversal of these effects with isoproterenol also appeared to be attenuated.

ANTIFIBRINOLYTIC AGENTS

Cardiopulmonary bypass results in significant fibrinolysis and several agents are commonly used to attempt to limit this effect and reduce bleeding. E-aminocaproic acid (EACA) and tranexamic acid (TA) are both carboxylic acid derivatives that are structurally related to the amino acid lysine [25] . Previous studies have determined that both EACA and TA are effective at reducing bleeding and transfusion requirements in infants and children undergoing cardiac surgery with bypass, particularly those who are either undergoing repeat stemotomy or who are cyanotic [26,27] . Aprotinin is a serine protease inhibitor isolated from bovine lung. Its ability to inhibit trypsin, plasmin, and kallikrein has been recognized since the 1930s, with clinical use first reported in 1953 for the treatment of acute pancreatitis [28,29] . At plasma levels of only 50 kallikrein inhibitor units (KIU)/mL, aprotinin is able to inhibit plasmin. At higher plasma levels (200 KIU/mL), aprotinin inhibits kallikrein formation, thus attenuating its central role in the production of post CPB coagulopathies and the activation of the inflammatory response [30] .

RECOMBINANT FACTOR VIIA

Recombinant factor VIIa (rFVIIA) was originally approved for use in hemophilia patients who had developed inhibitors to factors VIII or IX. In doses of 90μg/kg it was shown to be effective at treating bleeding in these patients [31].

FRESH VERSUS RECONSTITUTED WHOLE BLOOD

Debate continues as to whether fresh whole blood (< 48 hr) reduces bleeding and transfusion requirements, particularly in infants. The classic study by Manno and coworkers [32] demonstrated that either very fresh whole blood (< 6 hr), or fresh whole blood (24-48 hr) reduced blood loss by 85% in patients less than 2 yr of age who were undergoing cardiac surgery with bypass, when compared with reconstituted whole blood. A recent prospective, randomized, controlled study of 48 hr old whole blood versus reconstituted whole blood in 200 infants under 1 yr of age, demonstrated similar blood loss, blood transfusion requirements, and serum levels of inflammatory mediators and troponin I in both groups. The group received 48 hr old whole blood averaged a longer intensive care unit stay by one day, and postoperatively had a larger positive fluid balance [33].

BRAIN MONITORING

Currently the following brain monitors are in common use during cardiac surgery: processed electroencephalogram (EEG), transcranial Doppler ultrasound (TCD), and near-infrared cerebral oximetry (NIRS).

EEG

The Bispectral Index (BIS) monitor uses a proprietary algorithm based on the normal adult EEG; Fourier transformation and bispectral analysis of a one-channel processed EEG pattern compute a single number, the BIS. This index ranges from 0 (isoelectric EEG) to 100 (awake). Mean awake values in are the 90 – 100 range in adults, infants and children. The real-time unprocessed EEG waveform of the BIS can be used to recognize EEG burst suppression, or electrical silence.

TCD

TCD is sensitive, real-time monitor of cerebral blood flow velocity and emboli during congenital heart surgery. Currently available instruments use pulsed-wave ultrasound at 2 MHz frequency, which is range-gated, emits a power of 100 mW, and has a sample volume length of up to 15 mm. a display of the frequency spectrum of Doppler signals is easily interpreted, and peak systolic and mean flow velocities, in cm/sec, are displayed. The temporal or anterior fontanelle windows (in infants) are insonated at a depth sufficient to obtain a signal from the middle or anterior cerebral arteries on the side of interest.

NIRS

NIRS is a non-invasive optical technique used to monitor brain tissue oxygenation. Most devises use 2 – 4 wavelengths of near-infrared light at 700 – 1000 nm, where the iron-porphyrin complexes of oxygenated and deoxygenated hemoglobin have distinct absorption spectra. Commercially available devices measure the concentration of oxy – and deoxyhemoglobin, thereby determining cerebral oxygen saturation. The INVOS 5100 is FDA approved for use in children, is compact, simple to use, and requires no warm up. It uses two near-infrared wavelengths of 730 and 810 nm. The INVOS processor displays a numerical value, the regional cerebral oxygen saturation index (rSO_2i), which is the ratio of oxygemoglobin to total hemoglobin in the light path. The rSO_2i is reported as a percentage on a scale ranging from 15% to 95%. Clinical evidence suggests a correlation between low cerebral saturation and adverse neurological outcome.

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