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Mr. Speice

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Research Assessment 4

Subject: Congenital Heart Defects and Anaesthesia

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Assessment:

For most of my Independent Study and Mentorship II journey I have chosen to research tissue engineering and challenges specific to pediatric cardiothoracic surgeons. With a goal to better understand the surgical process and the pediatric cardiothoracic surgical team dynamic, I have chosen to research challenges faced by anaesthesiologist in pediatric cardiothoracic surgery in the hopes to gain a deeper understanding of anaesthesiology for my interview with Dr. Luis Zabala at Children's Health Dallas.

This article provided mostly background information on congenital heart defects. Most information presented in this background was very broad and most of what I had researched during my Independent Study and Mentorship I journey. However, it spoke of the various processes of anaesthesiology during non-surgery catheterization. Since being under anaesthesia can bring risk to pediatric patients it is important that medical professionals avoid putting a child

under anaesthesia more than once. This article also spoke of challenges related to specific congenital heart defects. Specific defects, age, and size can increase the risk of surgery.

Although not the best article due to its broad material it highlighted the importance that risk plays in pediatric cardiothoracic surgery. It is important for every professional in the pediatric cardiothoracic surgical team to take preventative measures to ensure that each patient is has the least possible risk of intraoperative and postoperative complication. This is seen in the procedures in the OR, surgeon preferences in tissue grafts, CVICU critical care rounds, and many other careers in the Heart Center. During my research on Critical Care in pediatric cardiothoracic surgery in my Original Work project for my Independent Study and Mentorship I journey I saw the unpredictability of the post operative care of pediatric cardiac patients. This article tied that idea by highlighting the different risks and preventative measures taken by anaesthesiologist preoperatively and intraoperatively. By better understanding the preventative measures taken by the Heart Center team I am able to better understand the challenges faced by the team and the medical innovations that allow for the prognosis of patients suffering congenital heart defects to improve.

With a better understanding of anaesthesiology in pediatric cardiothoracic surgery I am now able to raise more knowledgeable questions to Dr. Zabala and gain a lot more from my interview. Articles like these raise questions of improvement and innovation in pediatric cardiothoracic surgery, and by learning the challenges and complications faced by different physicians on the Heart Team and how they are faced today I can become one step closer of possibly developing improvement and innovation ideas of my own. Continuing research like this in my Independent Study and Mentorship program will grow my understanding of my topic and will lead to a possible final product idea.

Overall, I hope to use this assessment to create knowledgeable questions to Dr. Zabala that will broaden my understanding of the challenges he faces during surgery and recent innovations that have improved the challenges he faces. This assessment will continue to be a part of my search for an original work or final product.

Congenital heart diseases and anaesthesia

Sandip Junghare and Vinayak Desurkar

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Patients with congenital heart diseases (CHDs) are at increased risk of developing complications during anaesthesia. Improvements in medical and surgical management in recent decades have resulted in significantly more children with CHD surviving to adulthood. The aim of this article is to focus on broad classification of CHD and to provide an updated review on the current perioperative anaesthetic management of CHD patients in different settings such as (a) interventional cardiac procedures that have dominated the field, (b) uncorrected patients for non-cardiac surgery and (c) corrected patients for non-cardiac surgery. The complexity of the defects along with a variety of non-cardiac surgery makes it impossible to have one single-anaesthesia technique. Search on Ovid, PubMed, Google Scholar and Medline were done with MeSH terms such as 'congenital heart disease', 'cardiac catheterisation', 'anaesthetic management' and 'non-cardiac surgery' mainly focusing on review articles and controlled studies for preparing the article.

Introduction

Congenital heart disease (CHD) is a structural and functional heart disease, which is present at birth. Incidence of CHD is about 8–10/1000 live births worldwide and varies with modern diagnostics.[sup][1] We have no community-based data for the incidence of CHD at birth in India as a large number of births in India are not reported. The prevalence of CHD in India reported in 2005 was around 2.5–5.2/1000 live births, and common lesions were ventricular septal defect (VSD), patent ductus arteriosus (PDA), transposition of great arteries (TGA) and pulmonary atresia.[sup][2] A study conducted between 2011 and 2014 showed the prevalence in India to be as high as 19.4/1000 live births. Common CHDs were VSD (33%), atrial septal defect (ASD-19%) and tetralogy of

Comment [1]: This article will assist me in better understanding the challenges faced by a pediatric cardiothoracic anaesthesiologist, and will prepare me for my interview with Dr. Zabala on September 26th.

Comment [2]: one of the most prevalent factors of infant mortaity

Fallot (ToF-16%) in the age group of 0–5 years. In adults, it was 2.4/1000 with ASD being the most common defect.[sup][3] In 10-15% patients, surgical intervention may be required for associated extracardiac anomalies (airway, skeletal, genitourinary and gastrointestinal). Improvements in medical and surgical management have resulted in significantly more children with CHD surviving into adulthood. Anaesthesia-related cardiac arrest during non-cardiac surgery is more common in these patients.[sup][4] For uneventful anaesthesia in these patients, we need to understand the physiology and recognise the associated risks.[sup][5],[6] The complexity of the defects along with a variety of non-cardiac surgical procedures makes it impossible to have one single-anaesthesia technique. This review article emphasises the early recognition of the risk, understanding the physiology, advantages of a multidisciplinary approach and utility of newer modalities in anaesthetic management of these patients. It will focus on anaesthesia for diagnostic and therapeutic cardiac catheterisation, uncorrected CHD for non-cardiac surgery, grown up congenital heart disease (GUCHD) and adult patients with corrected congenital cardiac lesions presenting for non-cardiac surgery. Ovid, PubMed, Google Scholar and Medline were searched with MeSH terms 'congenital heart disease', 'cardiac catheterisation', 'anaesthetic management' and 'non-cardiac surgery', mainly focusing on review articles and controlled studies.

Comment [3]: From my experiences at Children's Health most children have a great prognosis if treated early.

Comment [4]: all about prevention

Classification of Congenital Heart Disease

Congenital heart disease is broadly classified as a) cyanotic and acyanotic CHD and b) conditions with shunt and without shunt[sup][7] [Table 1].{Table 1}

Recent advances in cardiac surgery and availability of newer palliative procedures have contributed to increased survival of patients with cyanotic heart disease and with single ventricle. This has resulted in subset of patients who have undergone corrective or palliative surgery, and have some limitations, including: (a) Blalock-Taussig (BT) shunt, (b) Norwood procedure, (c) Fontan procedure with single ventricle, (d) TGA with complete correction, (e) intracardiac repair for double outlet right ventricle (DORV) and ToF, (f) corrected total anomalous pulmonary venous connection (TAPVC) and (g) corrected with device closure or stenting.

Physiology

Cardiac surgery can correct CHD by way of either biventricular repair (complete repair) or univentricular repair (palliative). It is important to know the physiology of circulation in patients with CHD[sup][5] that can be divided into (A) normal circulation or series circulation, (B) parallel or balanced circulation and (C) single ventricle circulation.

Normal circulation

The systemic and pulmonary circulations work together in series. Most types of repaired CHD such as ASD, VSD and ASO (Arterial Switch Operation) have this type of circulation. Occasionally, surgeons deliberately keep fenestrations, similar to patent foramen ovale (PFO) or small VSD as a pressure release mechanism, which leads to some shunting. Left-to-right shunts result in increased pulmonary blood flow (PBF) and potentially decreased systemic blood flow; right-to-left shunts cause deoxygenated blood to flow into the systemic circulation, causing cyanosis and reduced PBF. Changes in systemic vascular resistance (SVR) and pulmonary vascular resistance (PVR) as a result of anaesthesia, including the administration of oxygen, impact on the behaviour of the shunt depending on its size. Large, unrestricted defects such as a large VSD may exhibit 'balanced' circulation physiology.

Balanced (parallel) circulation

Here, the systemic and pulmonary circulations are connected by some means and physiologically work in parallel. Connections can be natural defects such as VSD, PDA or ASD. Artificial connections are uncorrected TGA with balloon atrial septostomy (ASD), modified BT shunt and PDA stents. In these patients, pulmonary and systemic blood flow is balanced with SVR and PVR. These groups are at risk because anaesthetic drugs can cause changes in SVR and PVR, resulting in unbalancing of PBF. High pulmonary blood flow leads to pulmonary oedema and desaturation and reduced systemic perfusion. Lower PBF leads to desaturation and acidosis.

Single ventricle physiology

Full anatomic correction (biventricular repair) is not possible in some congenital defects such as hypoplastic left heart syndrome or double-outlet right ventricle. In single ventricle physiology, only one ventricle works as a systemic ventricle and the other is rudimentary. The pulmonary blood flow is passive based on pressure

Comment [5]: multiple CHDs

Comment [6]: challenge

Comment [7]: broad term

Comment [8]: research this

gradient between the pulmonary artery (PA) and left atrium. Usually, these patients have three-staged surgical palliation; BT shunt or PA band in infancy; Glenn or hemi-Fontan (superior vena cava connected to PA) at 1[sup]st or 2[sup]nd year of life and then a Fontan procedure – both inferior and superior vena cava connected to PA. The pulmonary blood flow is passive and hence changes in PVR and positive intrathoracic pressures compromises PBF. However intermittent positive pressure ventilation (IPPV) may be needed to avoid hypercapnoea and hypoxia, and minimal peak inspiratory pressures and inspiratory times may optimise PBF.

Risk Assessment

A point system for the risk stratification of a CHD patient before undergoing a procedure was developed by Mossad.[sup][10] Children and adults with heart disease are at increased risk of mortality and morbidity when undergoing non-cardiac surgery.[sup][4],[9] Risk associated with an individual patient is based on several criteria [Table 2].[sup][4],[5],[8]{Table 2}

The patients at highest risk are infants with a functional single ventricle and patients with suprasystemic pulmonary hypertension (PHT), left ventricular outflow tract obstruction, and cardiomyopathy.[sup][4],[11] The presence of long-term sequelae such as cardiac failure, PHT, arrhythmia and cyanosis indicates a complex problem. The usual procedural risks during various catheterisation laboratory interventions are coronary ischaemia, cardiac arrest, low cardiac output, RV failure, pulmonary hypertensive crisis, arrhythmias, cardiac perforation and tamponade.[sup][12]

Pre-Operative Assessment

Good preoperative assessment is essential to determine the physiological status of the patient. This includes recording the height and weight, thorough examination of the cardiovascular and respiratory systems and the presence of cyanosis, clubbing or squatting episodes.

Signs and symptoms of poor cardiac output and heart failure include difficulty in feeding, poor growth, sweating in infants or reduced exercise tolerance with fatigue in older children. Increased respiratory rate, chest retraction, nasal flaring, use of accessory muscles of respiration, pedal oedema, jugular venous distention, enlarged liver and rales suggest cardiac failure.

Peripheral pulses and blood pressure in all extremities should be measured (abnormal findings in coarctation of the aorta and BT shunt). Similarly, oxygen saturation by pulse oximetry should be measured in all limbs (differential cyanosis). Oxygen saturation after exercise can give some idea about heart function. Association with Down's syndrome is common and hence atlantoaxial subluxation should be kept in mind. The child with history of prolonged intubation can have subglottic stenosis. Many patients, especially adults, can have implanted pacemakers and/or automated defibrillators. The current medications should be reviewed and administered unless there are any contraindications on the day of surgery [Table 3].{Table 3}

Cardiology Review Before Procedure

Need for a cardiology review depends on the complexity of the lesion. CHD patients who have had complete repair do not need a cardiology reference if they are fit and healthy. A standard pre-anaesthetic visit without cardiology consultation is acceptable. For complex lesions and major surgeries, cardiology reference is advocated, and especially if patient's condition has changed recently. However, clearance must always be given by an anaesthesiologist as the cardiologist will not have full knowledge of anaesthetic effects and surgical procedures.[sup][13]

In adult patients coming to surgery, one should look for long-term problems that vary with the disease condition [Table 4].[sup][14],[15]{Table 4}

Pulmonary Hypertension and Eisenmenger Syndrome

Left-to-right shunt causes increased pulmonary blood flow. The amount of flow determines the response of pulmonary vasculature. In the initial period, the pulmonary vasculature will accommodate the flow (unless heart failure occurs due to left ventricular overload e.g., large PDA or VSD). Persistent exposure of the pulmonary vasculature to increased blood flow, as well as increased pressure, may result in pulmonary arteriopathy (muscular hypertrophy) which leads to increased pulmonary vascular resistance with mean PA pressure >25 mmHg at rest or >30 mmHg with exercise. The pulmonary capillary wedge pressure (PCWP) is ≤15 mmHg. This PHT[sup][8] can present as follows:

*Dynamic - related to high shunt flows that respond to reduction of the shunt *Reactive - is the difficult variety, and challenging to control in perioperative periods *Shunt Reversal-Eisenmenger Physiology.

The first variety can be part of balanced circulation physiology and should be looked after during anaesthetic management of left-to-right shunts. Reactive PHT occurs in older children or adults with untreated shunts. It may be still responsive to oxygen, but also increases with stimuli that cause pulmonary vasoconstriction.[sup][11] Avoiding sympathetic stimulation and use of nitric oxide can be lifesaving in these patients [Figure 1].{Figure 1}

Eisenmenger syndrome is shunt reversal due to suprasystemic pulmonary arterial pressures and conversion of acyanotic left-to-right shunt to cyanotic right to left shunt. Generally, atrial level shunts will take more time than ventricular level shunts for development of pulmonary arterial hypertension. Adult patients with Eisenmenger syndrome are most challenging for anaesthetic management. The signs and symptoms in the advanced stages include central cyanosis, dyspnoea, fatigue, haemoptysis, syncope and right-sided heart failure.

As a consequence of chronic slow progressive hypoxaemia with central cyanosis, adult patients suffer from multiple system problems including coagulation disorders (bleeding complications and paradoxical embolism), renal dysfunction, hypertrophic osteoarthropathy, heart failure, reduced quality of life and premature death. Iron deficiency should be addressed in these patients as it is one of the strongest independent predictors of thrombosis.

For a long time, therapy has been limited to symptomatic options or lung or combined heart–lung transplantation. New selective pulmonary vasodilators have become available and proven to be beneficial in various forms of pulmonary arterial hypertension. Drugs such as bosentan and sildenafil are being used and this targeted medical treatment has been expected to show promising effects with a delay in deterioration, including patients with Eisenmenger syndrome.

Pre-Medication

Good pre-medication is important to reduce anxiety and make parental separation easy, which in turn will help in smooth induction. This can reduce catecholamine release and avoid hypercyanotic spells in children with Fallot's

Tetralogy. Hypoventilation, hypercarbia lead to pulmonary hypertension, and must be avoided, and pulse oximetry monitored after giving pre-medication. Choices of drugs include: midazolam up to 0.5 mg/kg orally (up to a maximum 20 mg) or 0.05–0.2 mg/kg intravenous (IV), Triclofos (pedicloryl) oral 50–75 mg/kg half an hour before the procedure, or Ketamine (1–2 mg/kg IV or 5 mg/kg oral if IV access is absent).

Endocarditis Prophylaxis

The American Heart Association has advised the use of antibiotics only in “high-risk” patients (before dental procedures):[sup][8]

*Patients with prosthetic cardiac valves *Patients with prior infective endocarditis *Patients with unrepaired or palliated cyanotic CHD including shunts and conduits *Patients with CHD repair with prosthetic material or device placed by surgery or catheter intervention during first 6 months after placement *Patients with CHD repair with residual defect at the site or adjacent to the site of prosthetic patch or device that inhibits endothelialisation.

Endoscopic procedures need not have any prophylaxis.

Anaesthesia Management

Standard pre-operative fasting guidelines should be followed, keeping in mind dehydration, high haematocrit and the need for adequate preload.[sup][12]

Appropriate monitors should be applied before induction of anaesthesia if the child is cooperative. Intravenous (IV) or inhalation induction may be carried out, depending on the availability of (IV) access, and the child's physiological condition and cooperation.

Shunt flow behaviour depending on various events is shown in [Figure 2]. This affects pulmonary or systemic blood flow, which can have impact on cardiac output and perfusion.[sup][16]{Figure 2}

The anaesthetic management is summarised in [Table 5] depending on the type of surgery and physiology, and effects and doses of drugs in [Table 6].[sup][19]{Table 5}{Table 6}

Sevoflurane is agent of choice for inhalation induction. Propofol or ketamine are used for IV induction. The likely physiological consequences of varying systemic and pulmonary vascular resistances on shunts and cardiac output must be considered. Tracheal intubation is required for the majority of cases, especially

neonates and, infants, and is facilitated with a neuromuscular blocking agent (e.g., atracurium 0.5 mg/kg). Older children undergoing short procedures may occasionally be managed using a supraglottic airway device. The airway should be controlled during procedures associated with high risk of peri-procedure haemodynamic instability, procedures with high risk of complications, patients in whom internal jugular venous access is required or who may require resuscitation.

Due to use of transoesophageal echocardiography during interventional procedures, tracheal intubation is essential in children. Adults can be managed under sedation and local anaesthesia.

Maintenance

During the catheterisation laboratory procedures, it is essential to keep the patient immobile, maintain haemodynamics as close to pre-procedural values as possible in addition to maintaining normothermia and normocapnia. Stable haemodynamics are required to generate meaningful baseline pressures and to allow interpretation of diagnostic interventions such as stress testing and nitric oxide without confounding factors. High inspired oxygen concentrations (>30%) may give erroneous results in flow studies and may decrease pulmonary vascular resistance, thereby increasing left-to-right shunt fraction. Oxygen and air with an inhalation agent are the preferred method for maintenance of anaesthesia. Increased inspired oxygen concentrations are used when attempting to reduce pulmonary vascular resistance in conjunction with inhaled nitric oxide, when investigating pulmonary hypertension.

Intraoperatively, only small doses of fentanyl (1–2 [micro]g/kg) are required to blunt haemodynamic changes during stimuli such as insertion of femoral sheaths or transoesophageal echocardiography probes. Paracetamol and local anaesthetic infiltration are usually adequate for post-procedural analgesia. An iv antiemetic (dexamethasone 0.2–0.5 mg/kg or, ondansetron 0.1 mg/kg) is usually given to avoid nausea and vomiting.

Isotonic maintenance fluids will be required in the vast majority of cases, with attention to blood sugar monitoring in neonates. It is important to account for the volume and content of flushes and IV contrast used by the operator and also blood loss, both of which may be considerable. Volume loading can impair

cardiac function. Iodinated contrast media have some nephrotoxic potential. Risk factors include pre-existing renal impairment, diabetes, heart failure and use of other nephrotoxic drugs; however, problems can still occur in patients with previously normal kidneys. Dehydration should be avoided, other nephrotoxic drugs omitted and where risk is high, minimum volumes of iso-osmolar or low osmolar contrast medium are used.

The majority of patients are extubated at the end of the procedure and recovered in a routine fashion, with special attention to the femoral puncture sites and lower limb perfusion. Paediatric/adult intensive care is reserved for ill or higher risk cases, those with pulmonary hypertension and those where serious complications have occurred.

Post-operative care should be given by experienced staff caring for this subset of adult and paediatric patients. Good pain relief and control of nausea-vomiting along with the specific events in these groups of patients such as dysrhythmias, bleeding and thromboembolic events should be addressed.[sup][16]

Complications after catheterisation laboratory procedures after sedation or general anaesthesia, are airway events (bronchospasm, laryngospasm, aspiration and apnoea), cardiovascular events (hypotension, arrest and arrhythmias) and post-operative events (nausea, vomiting, emergence agitation, apnoea and hypoxia).[sup][19] Respiratory events are more common in infants and intubated patients.[sup][12] Risk of adverse events is highest with neonates and infants, followed by children and then GUCHD adults. Risk of cardiac arrest is more during interventional procedures such as VSD device closure, in neonates, and can be attributed to stiff wires and catheters inducing arrhythmias[sup][20],[21] [Table 7].{Table 7}

Newer Modalities of Surgical Treatments in Patients With Congenital Heart Disease

Laparoscopic and video-assisted surgery is now becoming standard practice in general, gynaecological, urological and thoracic surgery. Inflation of CO[sub]2 into cavities can be physiologically challenging during procedure. Positioning (head down, lateral, prone) can have dramatic effects with IPPV, especially in preload dependent circulation. Intraoperative invasive monitoring of blood pressure and central venous pressure (CVP) along with blood gas

measurements is warranted in these cases in GUCH patients. Closed procedures will have to be converted to open if situation worsens and that should be a planned event.[sup][22]

Airway surgery can be common in patients with GUCH due to prolonged intubations with need for rigid bronchoscopy or suspension laryngoscopy for glottic and subglottic region procedures. Maintenance of oxygenation and physiological goals remains the main stay of management.

For post-operative pain relief, regional blocks, epidurals both lumbar and thoracic can be used keeping in mind the coagulation issues. IV patient-controlled analgesia (PCA) with narcotics can be challenging in those with reactive pulmonary pressures in response to rise in PaCO[sub]2 that might occur with opioid PCA.

Pregnancy and corrected or uncorrected CHD is another challenging group of patients, especially those with Eisenmengers syndrome. Corrected heart disease patients will behave as normal unless their cardiac function is moderately impaired before pregnancy. Uncorrected patients with a balanced circulation or single ventricle physiology will need judicious application of knowledge of pathophysiology and effects of anaesthesia. There is no single technique to apply; rather patient based decision making is important. Use of regional anaesthesia has been effectively used in such circumstances supplemented with vasopressors if needed.

Anaesthetising patients with CHD is challenging and there are no evidence-based recommendations for management. Given the scope of abnormalities and advancing treatment options, it is difficult to propose any single approach and hence multidisciplinary approach involving anaesthesiologists, surgeons, cardiologists, intensivists, paediatricians and neonatologist is essential in decision-making process.

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Conflicts of interest

There are no conflicts of interest.

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