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Anaesthetic Considerations for Congenital Heart Disease Patient

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1. Introduction

Incidence of congenital heart disease (CHD) is about 0.8%[1] and most of these CHD children (80%) survive to adulthood in developed countries due to early diagnosis and intervention along with improved surgical and anaesthetic techniques. But the situation is different in most of the third world countries, where 90% of these children receive suboptimal or no care[2]. These patients commonly admitted in the hospital for procedures like cardiac catheterization, radiological procedures[3 4], dental and cardiac surgery.

There is increased risk of mortality and morbidity[5] under anaesthesia as their anaesthetic management in the operating room is challenging in several respect. Few heart defects are so complex that you need to involve paediatric cardiologist and intensivist for complete understanding of anatomy and pathophysiology of heart defect.

Adult population with congenital heart defects has also increased[6 7] over the years and poses more challenges for anaesthesiologist in perioperative period. It is now expected that soon there will be more adult with congenital heart defects than children. Grown up congenital heart (GUCH) is a separate entity, which requires expertise of different disciplines to prevent associated morbidity and mortality[8] during operations (cardiac or non cardiac) particularly in uncorrected defects and in pregnant patients.

When a cardiac defect is recognized in a paediatric patient then the presence of other cardiac and extracardiac lesion is a possibility. The incidence of extra cardiac malformation is as high as 20 – 45% and chromosomal abnormalities in these CHD patients is found to be 5-10%.

Perioperative anaesthetic considerations include preoperative evaluation, management of hypoxaemia, shunt, polycythaemia, pulmonary hypertension and ventricular dysfunction.

2. Classification

Several classifications of CHD have been introduced. Two are given below

2.1 Cyanotic/Acyanotic CHD

2.1.1 Acyanotic

a. Ventricular Septal Defect (VSD)
b. Atrial Septal Defect (ASD)
c. Patent Ductus Arteriosus (PDA)
d. Atrio ventricular Septal Defect (AVSD)

e. Pulmonary Stenosis (PS)
f. Aortic Stenosis (AS)
g. Coarctation of the Aorta

2.1.2 Cyanotic CHD

a. Tetralogy of Fallot
b. Transposition of the Great Arteries (TGA)
c. Total Anomalous Pulmonary Venous Return(TAPVR)
d. Tricuspid Atresia
e. Truncus Arteriosus
f. Uncommon, each <1% of CHD, pulmonary atresia, Ebstein's anomaly.

2.2 Classification on the basis of pulmonary and systemic flow

1. Excessive Pulmonary Blood Flow
   a. VSD, ASD, PDA, PAPVR
2. Inadequate Pulmonary Blood Flow
   a. Tetralogy of Fallot, Pulmonary atresia
3. Inadequate or obstruction to systemic blood flow
   a. Coarctation of Aorta
4. Abnormal Mixing
   a. TGA

3. Preoperative consideration

Three type of paediatric CHD patients are expected to come for evaluation.

1. Patients with uncorrected cardiac defect
2. Patients who had previous palliative surgery
   a. ToF with BT shunt
   b. Atrial septostomy for TGA
3. Patients in whom total correction has been done but they may have residual defects requiring certain procedures[9]

Preoperative evaluation should include detailed information about cardiac lesion, altered physiology and its implications. There are few questions which should be clearly answered during preoperative evaluation of these CHD patients. These includes

a. Complete understanding of the anatomical changes due to cardiac defect or palliative procedure
b. Direction and amount of shunting
c. Presence and severity of pulmonary hypertension
d. To what extent pulmonary flow reduced or increased?
e. Degree of hypoxaemia, Polycythaemia
f. Coagulation abnormalities
g. What associated pathophysiological findings likely will influence the management?
h. Functional status of the patient

Fatigue, headache, visual disturbances, depressed mentation and paraesthesia of toes and fingers are presenting symptoms of polycythaemia. History of cyanosis and congestive heart failure (CHF) are major manifestations of CHD. Fatigue and dyspnoea on feeding and irritability indicate poor functional status. Cyanosis occurs due to decrease pulmonary flow

anatomically or functionally (Mixing lesion). Cyanosis may be permanent or appear intermittently. Cyanosis may not be seen in new born due to presence of faetal haemoglobin which is highly saturated at a given PaO2.

High pulmonary flow leads to pulmonary edema. Failure to thrive and feeding problems are common in patients with history of repeated pulmonary congestion. Patient may presents with tachycardia, tachypnoea, irritability, cardiomegaly and hepatomegaly. The right ventricular function should also be assessed as it is equally important in paediatric CHD patient.

Try to avoid dehydration in cyanotic CHD patients by allowing clear liquids two hours prior to surgery (Table 1). Children also have low glycogen stores which makes them vulnerable to hypoglycemia. If timing of surgery uncertain then start an intravenous line and give glucose containing solution. Midazolam[10 11] is a preferred sedative in the doses of 0.5 to 1mg/kg or even higher doses in few studies given orally half hour before surgery (Table 2). If patient is on prostaglandin (PGE1) infusion then it should be continued.

2hrs	Clear liquids (water, apple juice, pedialyte)
4hrs	Breast milk
6hrs	Formula & Cow milk
6hrs	Solids

Table 1. NPO Orders

Age	**Premedication**
< 6 months	None
6 months to 8 years	Midazolam Oral 0.5- 1.0mg/kg (Max. 12 mg) Intravenously 0.05 – 0.2mg/kg Chloral hydrate 40 – 50mg/kg
> 8 years	Midazolam 7.5 mg PO Morphine 0.1mg/kg IM Ketamine 4mg/kg IM

Table 2. Premedication orders

4. Investigation

Polycythaemia is very common which increases blood viscosity and leads to thrombosis and infarction in cerebral, renal and pulmonary region. Although polycythaemia leads to intravascular volume expansion but at the same time reduces plasma volume. Coagulation abnormalities also occur due to hypofibrinogenaemia and factor deficiencies. Platelet count, PT and PTT should be ordered in all patients coming for surgery. Preoperative phlebotomy can be performed in patients with symptomatic hyperviscosity and haematicrit > 65%.

Electrolyte abnormalities are commonly seen in patients who receive diuretics and parental nutrition. Hypocalcaemia commonly found in patients with Di George syndrome.
ECG may show ventricular strain or hypertrophy pattern. Echocardiography is used for doppler and color flow mapping while catheterization is used for information about pressures in different chambers, magnitude of shunt and coronary anatomy. Examine chest X-Ray for heart position (Dextrocardia) and size, atelectasis, acute respiratory infection, vascular markings and elevated hemidiaphragm. High pulmonary flow will leads to increased pulmonary marking while reduced flow causes oligaemic lung fields.
Neurological assessment and MRI[12] may also be needed in these patients. Delayed brain development is associated with certain CHD. Fetal MRI can help in early assessment of immature brain.

5. Intraoperative considerations

Presence of CHD in paediatric patients poses a great challenge for anaesthetist[13] as morbidity and mortality is quite high. Incidence of cardiac arrest in these paediatric patients under anaesthesia is higher[14] than non CHD patients and mainly due to pharmacological interaction and over dose[5].
Intravenous line must be placed in all patients even for minor procedure. All intravenous tubings should be free of air bubble. Polycythaemic patient must be well hydrated before induction either by IV or orally.
Sevoflurane[15] is preferred over halothane due to better haemodynamic stability in CHD patients. Most of the CHD patients tolerate inhalation induction with sevoflurane while patients with poor cardiac function, may not tolerate inhalation induction. Ionotropes should be continued if patient is on ionotropes.

5.1 Monitoring

Monitoring in paediatric CHD is the same as in adult cardiac surgery but there are few differences and considerations during surgery. Monitoring during surgery ranges from simple ECG to blood glucose, which is controversial due to non availability of evidence that tight blood sugar control improves outcome[16].

5.1.1 Electrocardiogram

Although ECG can be helpful in the detection of ST changes but is mainly used for arrhythmia detection in paediatric patients. Even arrhythmia detection is difficult due to baseline tachycardia. Skin should be prepared for electrode by rubbing with alcohol pad or swab. Three leads system is commonly used while in older children five leads system can also be used.

5.1.2 Blood pressure monitoring

Non invasive monitoring

Non invasive blood pressure should always be monitored even in the presence of arterial line. Cuff should be 20% wider than the diameter of limb where non invasive blood pressure is monitored. Smaller cuff results in erroneously high pressure while larger cuff will give lower pressures.

Invasive pressure monitoring

It not only provides beat to beat continuous blood pressure monitoring but also provides easy access for blood sampling. Pressure monitoring tubing and stopcocks should be free of air to prevent air embolism and damping of system. It is also a major source of fluid overload as system continuously flushes 2-4 ml/hr per invasive line. In addition a quick flush also pushes about 1-2 ml of fluid per second . Dextrose can be used but usually normal saline is the flushing solution as bacterial growth is less likely.

5.1.3 Central venous pressure

Central venous access not only helpful in monitoring but also provides a reliable route for drugs, fluid and blood. Right internal jugular vein (Table 3) is commonly used due to its straight course to right atrium while left side is avoided due to concerns about its persistent connection to left SVC (which may be ligated during surgery). Alternatively femoral and subclavian veins can also be used.

Weight	CVP
< 5kg or less than one year	4F, 5cm
5 – 20 kg	4F, 8cm
>20 kg	5F, 8 or 13cm
Adult	7F

Table 3. Central venous catheter (Internal Jugular Vein) size according to weight

5.1.4 Pulse oximeters

Usually two oximeters are placed , one in the upper limb and other in the lower extremity. Pulse oximeter uses two light emitting diodes and one photodiode for detection of red and infra red lights.

Accuracy of pulse oximeter is affected by

1. Hypotension
2. Hypothermia
3. Electrocautery
4. Artifacts due to
    a. Thick skin
    b. Dark color
    c. Bright outside light
    d. Presence of dyes like indocyanine green and methylene blue
5. Abnormal haemoglobins
    a. Met Hb
    b. Carboxy Hb

Not affected by fetal hemoglobin.

5.1.5 Cerebral oximeter

Transcranial near-infrared spectroscopy (NIRS)[17] is a sensitive measure of regional hypoperfusion. It measures all haemogloblins and useful in non pulsatile cardiopulmonary bypass and circulatory arrest. Cerebral oximeter detects intravascular haemoglobin oxygen saturation of cerebral cortex.

5.1.6 Echocardiography

Intraoperative transesophageal echocardiography (TEE) plays a critical role in improving surgical outcome in CHD surgeries by confirming diagnosis and identifying residual defects. It is also helpful in the placement of devices in catheterization lab. Micromultiplane TEE probe and three dimensional technologies are new advances in echocardiography. Epicardial echocardiography is an alternative option in institutions where smaller TEE probe is not available[18]. Adult TEE probe can be used in patient weighing more than 20Kg.

Arterial blood gases and blood glucose should also be done frequently. Tight blood glucose control is suggested by certain authors as high blood sugar is toxic to mitochondria.

Intraoperative management

Anaesthetic management during surgery depends on presence or absence of shunt, pulmonary hypertension, hypoxaemia, Ventricular dysfunction, pulmonary flow and arrhythmia.

5.2 Shunt

Shunting through these defects depends upon diameter of defect and balance between systemic and vascular resistance. Balance between systemic vascular resistance (SVR) and pulmonary vascular resistance (PVR) is essential in the anaesthetic management of patient with shunts.

Normal pulmonary: Systemic ratio (Qp:Qs ratio) is 1:1 which indicate either no shunting or bidirectional shunt of equal magnitude. Qp:Qs ratio of 2:1 indicate left to right shunt while less than 1:1 ratio (0.8:1) means right to left shunt. The ratio is estimated from oxygen saturation measurements at pulmonary veins, pulmonary artery, systemic arterial and mixed venous blood.

5.2.1 Left to right shunt

1. Atrial septal defect (ASD)
2. Ventricular septal defect (VSD)
3. patent ductus arteriosus (PDA)
4. Atrio ventricular (AV) canal defects
5. Complete anamolous venous return (CAVR)
6. Partial anomalous venous return (PAVR)
7. Artificial Blalock taussig (BT shunt)

L –R shunt reduces greatly with drop in SVR or an increase in PVR. It leads to excess pulmonary blood flow. Patients are usually acynotic but deterioration in gas exchange may result from pulmonary congestion. Avoid 100% oxygen and hyperventilation in patients with L $\rightarrow$ R shunt.

Patients with PDA are vulnerable to coronary ischaemia[11] due to ongoing pulmonary runoff during the diastolic phase. Therefore diastolic blood pressure (DBP) should be monitored during surgery. Diameter of Modified Blalock Taussig shunt is fixed so its output is proportional to SVR and in case of systemic hypotension, the pulmonary blood flow will be reduced. Blood pressure in the arm will be low due to BT shunt, so use contralateral limb.

5.2.2 Right to left

These intra cardiac shunts lead to prolong inhalation induction. **R → L shunt** (e.g. tetralogy of fallot (TOF) or shunt reversal[12] occur when SVR drops or PVR increases. Hypercyanotic spell under anaesthesia will respond to volume, Increase SVR with alpha agonists such as Phenylephrine.

5.3 Hypoxaemia

Inadequate pulmonary blood flow and/or admixture of deoxygenated with oxygenated blood in systemic circulation are usually responsible for ischaemia. In addition pulmonary congestion with inadequate exchange of gases can also leads to hypoxaemia.
Persistent hypoxaemia leads to following changes

a. Slightly ↑ HR
b. Hyperventilation
c. Polycythaemia
d. Chemoreceptor response to hypoxaemia reduced
e. Cerebral and myocardial oxygenation maintain but Visceral and muscular oxygenation reduced
f. Reduced metabolic activity of many organs
    a. Growth retardation
g. Myocardial ischaemia can occur
h. Myocardial dysfunction
i. Down regulation of β - receptors

The anaesthetic management includes adequate hydration, maintenance of systemic blood pressure, minimizing additional resistance to pulmonary blood flow and avoids sudden increase in oxygen demand (crying, struggling, and inadequate level of anaesthesia).

5.4 Pulmonary hypertension (HTN)

During early stages, the pulmonary HTN is reactive and responds to hypothermia, stress, pain, acidosis, hypercarbia, hypoxia and elevated intrathoracic pressure but later pulmonary HTN becomes fixed. This last stage, where pulmonary vascular resistance exceeds SVR and symptoms appear due to R → L shunt, is the Eisenmenger syndrome[13].
Anaesthetic risk is quite high including right ventricular failure, bronchospasm, pulmonary hypertensive crisis and cardiac arrest. Anaesthetic management focus on preventing further increase in R-L shunt by keeping SVR high and PVR low, maintaining myocardial contractility and prevention of arrhythmia and hypovolemia.

5.5 Ventricular dysfunction

Chronic volume overload (Large shunts, valvular regurgitation), Obstructive conditions and cardiac muscle diseases leads to reduced ventricular function. Blood gas and X-Ray may

show metabolic acidosis and pulmonary edema respectively. Patients are usually on digoxin, diuretics and ionotropes.

Anaesthetic considerations includes

1. Preoperative optimization of following before surgery
   a. Ionotopes
   b. Diuretics
   c. Digoxin
   d. Antiarrhythmics or ablation in patients with arrhythmia
2. Preoperative CBC and electrolytes
3. Etomidate and fentanyl provide cardiovascular stability at the time of induction
4. Avoid or limit the use of inhalation anaesthetics due to associated myocardial depression
5. Maintain normal sinus rhythm
6. Maintain preload during anaesthesia.
7. After load reduction in certain situations

5.6 Miscellaneous concerns

5.6.1 Neurological outcome

There is growing concern about their quality of life and neurocognitive function, as the long term survival of these children is now possible. 20 -50% may develop neurological impairment due to chronic hypoxaemia, prolong deep hypothermic circulatory arrest and prolong exposure to anaesthetics. Non pulsatile low flow during cardiopulmonary bypass causing ischaemia/reperfusion injury may also play a part[19].

Brain adapts to chronic hypoxia due to presence of NMDA 2B receptors in early life. Cortical neurons may reduce by 30% due to chronic hypoxia causing reduction in brain volume. But this reduction is compensated when normoxia develops after surgery. Although most of the article have supported the use of high dose narcotics in over all outcome but at present there is no concrete evidence about best anaesthetic agents for congenital heart surgery.

5.6.2 Coagulation disturbances

Coagulation abnormalities are very common in CHD patients particularly in cyanosed and polycythaemic patient.

Coagulation derangement associated with polycythaemia includes:

1. Decreased platelet count and function
2. Primary fibrinolysis
3. Impaired coagulation factors production
4. Contracted serum volume

Use of blood products is common in paediatric cardiac surgery due to coagulopathy during surgery and several strategies have been instituted to minimize this practice. Preoperative exchange transfusion of 20 ml/kg FFP to replace same amount of blood is an effective method to counter coagulopathy. Antifibrinolytics like aprotinin and tranexamic acid[20] have been used for this purpose. Aprotinin is no longer recommended in cardiac surgery due to higher incidence of renal failure, stroke and myocardial infarction while the use of tranexamic acid has increased.

Tranexamic acid as a part of blood saving strategy is given as a bolus of 100mg/kg followed by 10 mg/kg/hr infusion. Whole blood transfusion is quite effective in coagulopathic

patients. Factor VII in the dose of 90 microgram/kg is increasingly used in paediatric congenital heart surgeries.

5.6.3 Grown up congenital heart (GUCH)

Pregnancy

Increased in blood volume during pregnancy may further aggravate the situation and patient may develop arrhythmias, pulmonary congestion and heart failure. Consideration during pregnancy ranges from termination of pregnancy to the safe delivery by caesarean section. A multidisciplinary approach involving obstetrician, paediatric cardiac surgeon, paediatric cardiologist, intensivist, anaesthetist and neonatologist is essential in decision making process.

Anaesthetic challenges and considerations include

1. Invasive line monitoring according to the severity of cardiac defect
2. Slow infusion of lowest effective dose of oxytocin as vigorous uterine contraction leads to high pre load
3. Use of inline air filter
4. Reduction in SVR should be avoided
5. Coagulation abnormalities should also be considered
6. Prevention of thromboembolic events

Eisenmenger

Most of the patients with Eisenmenger started with simple correctable cardiac defects but eventually leads to severe pulmonary hypertension (PVR > 800 dynes/cm^{-5}) which does not respond to pulmonary vasodilators. Hypoxaemia, myocardial dysfunction and arrhythmia is a common finding.

Perioperative risk includes

1. Arrhythmia
2. Cardiac arrest
3. Pulmonary hypertensive crisis
4. Bleeding
5. Thrombosis

Anaesthetic management includes

1. Phlebotomy in hyperviscosity syndrome
2. Avoid dehydration in preoperative period
1. Avoid myocardial depressants
2. Keep SVR high
3. Try to reduce PVR
4. Regional anaesthesia can be used but general anaesthesia is preferred
5. Postoperative pain should be adequately managed
6. Will require intensive care after surgery

6. Common CHD

6.1 Ventricular septal defect (VSD)

VSD is the most common congenital heart defect. It may be an isolated cardiac defect or may be associated with other cardiac defects like ASD, PDA or a part of complex defects

(tetralogy, AV canal defect). Communication between two ventricles can be of any size and can occur at any part of septum. Most common type of VSD is peri membranous (also called subaortic or infracristal). Other less common defects are subpulmonary (Supra cristal, infundibular or outlet type), Inlet type (canal type) and muscular. Spontaneous closure is possible in muscular and membranous type of defects.

Smaller defects are not associated with large shunting of blood from left ventricle to right ventricle may not diagnose early in life but they are prone to infective endocarditis. Whereas larger defects cause shunting of blood from left to right ventricle this led to higher pulmonary blood flow and consequently pulmonary congestion. Due to early development of symptoms these patients diagnosed earlier. During systole LV ejects blood not only in the aorta but also in the pulmonary artery causing volume overload of pulmonary vessels, atria and left ventricle. These patients will develop high pulmonary vascular resistance (PVR) and if untreated will leads to Eisemenger.

A device like amplatzer can be placed to close few of these defects by interventional cardiologist. This procedure is performed in the cath lab as a daycare procedure but there are certain criteria needs to be fulfilled. There should be an adequate rim around the defects where amplatzer can be placed. Surgically VSD can be approached through ventricle, aorta, pulmonary artery or right atrium.

Anaesthetic considerations

Always consider high pulmonary vascular resistance in these patients and be ready to treat high PVR and right ventricular failure by inhaled NO, dobutamine and milrinone. Desirable haemodynamic goals by anaesthetists are to have slightly higher preload and pulmonary vascular resistance while keeping the SVR on the lower side and at the same time maintaining heart rate and contractility. Up to 10% of patients may develop conduction abnormalities after VSD repair which may be transient or permanent.

Intraoperative transesophageal echocardiography (TEE) will be beneficial in recognizing residual defects, intracardiac air and right ventricular function. Smaller VSD are sometimes becomes apparent after closure of large defect. In uncomplicated VSD closure patient can be extubated in the operating room.

6.2 Atrial septal defect (ASD)

Normally there is no communication between right and left atria due to presence of a septum. This atrial septum composed of septum primum and septum secundum which merges with endocardial cushion, superior and inferior vena cava.

Several types of defects can occur in this septum leading to shunting across. Apart from secundum defect other less common are primum, sinus venosus and coronary sinus type.

Most common defect is **ostium secundum** which usually located in the centre (also called fossa ovalis type) and occurs due to deficient septum primum. It may be single or have several small defects called fenestrated type. Patent foramen ovale commonly seen at the same site in 25 – 30% of normal patients. Usually PFO do not permit left to right shunting but right to left shunting can occurs if right atrial pressure exceeds left atrial pressure (sneezing, valsalva)

Sinus venosus defect is usually associated with partial anomalous pulmonary venous drainage and appears either at the junction of superior vena cava and atrial septum (High up) or at the junction of inferior vena cava and septum (located lower part of septum). Repair some time may cause injury to SA node.

Ostium primum defect is due to failure of fusion between endocardial cushion and lower part of interatrial septum leading to communication between two atria and usually associated with cleft at anterior mitral leaflet.
Coronary sinus type defect is due to absence of wall between left atrium and coronary sinus leading to communication between left and right atrium. It may be associated with persistent left SVC.
Left to right shunting depends on the size of defects and compliance of ventricles as shunting usually occurs during diastole when both mitral and tricuspid valves are open. If the defect is small (less than 5 mm) then it's called restrictive type while larger defects are non restrictive and associated with right atrial dilatation, RV volume overload and increased pulmonary blood flow. Spontaneous closure is possible but most require device closure by cardiologist or surgery.

Anaesthetic considerations

Inhalation induction in infants and very young and intra venous induction in older children is acceptable technique. Intramuscular ketamine can be alternative for induction or intra venous line placement in some children. Pulmonary hypertension is generally not seen in these patients and their management is usually simple with the goals of higher preload and slightly high PVR to reduce pulmonary flow. Presence of ASD is not usually poses higher risk for infective endocarditis.
TEE is helpful to see the residual ASD, mitral valve repair (primum type), four pulmonary veins opening in left atrium (Sinus venosus type). Tracheal extubation in the operating room will help in minimizing the charges.

6.3 Tetralogy of Fallot (ToF)

ToF is the most common cyanotic CHD, accounting for 10% of all CHD. It comprises of four anatomical defects: (i) VSD (ii) RVOT obstruction (iii) RV hypertrophy (iv) Over riding of aorta.
VSD is usually large, non restrictive which led to equalization of RV and LV pressures and shunting through VSD depends primarily on systemic and pulmonary vascular resistance. RVOT obstruction is dynamic due to hypertrophied infundibulum but fixed obstruction can also occur due to v pulmonary valve stenosis.
Due to reduce pulmonary flow, main and branched pulmonary arteries hypoplasia may also be seen. Right ventricular hypertrophy is more marked when VSD is restrictive. ToF may also be associated with certain defects like anomalous origin of LAD crossing the RVOT, pulmonary atresia, absent pulmonary valve and complete AV canal defect.

Palliative surgery

The classic Blalock Taussig shunt was performed in 1944 to relieve the ToF related cyanosis where end to side anastomosis of subclavian artery to pulmonary artery was performed. Today modified BT shunt is the most commonly performed palliative procedure in CHD patients where a synthetic graft is interpositioned between subclavian artery and ipsilateral pulmonary artery.

Complete surgical repair

First total correction was performed by Lillehei in 1954. Surgical correction involves infundibular muscle resection through right ventriculotomy or transpulmonarry approach.

Pulmonary valve is removed or dilated accordingly and a transannular patch is placed. VSD is also closed at the same setting. Main Pulmonary artery and its branches are also inspected for narrowing. Some centres creates small ASD to counteract high right sided pressures. There is trend towards early total correction rather than palliative surgery which is followed by total correction.

Anaesthetic management

Goal of anaesthetic management is to avoid low SVR and ionotropes before bypass. If patient is on prostaglandin E1 then it should be continued in pre bypass period. Avoid catecholamine release in preoperative phase and at the time of induction by providing good premedication and adequate analgesia and anaesthesia.

Induction can be done with ketamine and fentanyl if intravenous line is in place. Inhalation induction can also be performed while maintaining SVR. Remember infundibular stenosis increased by increasing contractility and heart rate, so minimize noxious stimulus avoid catecholamine release. This is achieved by high dose fentanyl at the maintenance phase. Arterial line and central line should be placed after induction and intubation.

Acute desaturation at any time should be considered as tet spell and treated by analgesics and volume. Phenylephrine should also be available to treat low systemic vascular resistance and hypotension. Steroids given at the time of induction can help in reducing release of inflammatory markers during cardio pulmonary bypass.

TEE is helpful in assessing residual VSD and infundibular stenosis and degree of pulmonary regurgitation. In case of tet spell, give 100% O2, Phenyl ephrine, volume, increase depth of anaesthesia, hyperventilate and give bicarbonate. In addition esmolol or proponol can be tried to reduce infundibular spasm.

During postbypass period be ready for arrhythmias and heart block, RV dysfunction and coagulopathy. Ionotrpic support is mandatory in postbypass period along with high filling pressure particularly if right ventriculotomy was performed. Blood products should be available and antifibrinolytics should be started for coagulopathy.

6.4 Patent ductus arteriosus (PDA)

Ductus arteriosus is a normal communication in fetus, which constrict and closes within 10-15 hrs of birth and later closed anatomically by fibrosis in 2 – 3 weeks. Various mechanism have been described for initial functional closure, which includes increased PaO2, absence of placental derived prostaglandins and presence of catecholamines and bradykinins in new born.

Ductus venosus provides a communication between junction of main and left pulmonary artery and lesser curvature of descending aorta after left subclavian artery origin. Higher incidence of patent ductus arteriosus is seen in premature, females, children living at high altitude and associated with maternal rubella.

It provides left to right shunt causing high pulmonary flow and volume load on left atrium and ventricle. Pulmonary congestion and recurrent infection is commonly seen if remain open.

Medical management includes three doses of indomethacin. If medical management fails then either transcatheter or surgical closure becomes necessary. Surgical techniques include ligation via left thoracotmyn sternotomy or recently by video assisted thoracoscopy.

Anaesthetic considerations

Anaesthesia management is planned according to prematurity, degree of pulmonary congestion and PVR and surgical technique. During surgery aorta, left pulmonary artery and left main bronchus can be mistakenly ligated instead of ductus arteriosus. Remember to place a pulse oximeter at lower extremity to diagnose ligation of aorta. In addition, DBP will rise as soon as PDA is ligated, which will confirm the identification.

Invasive monitoring is not essential in uncomplicated PDA but arterial line can be placed in patients with comorbids to check beat to beat pressure and diagnosis and correction of acidosis. Limit Left to right shunt by keeping FiO_2 low and $PaCO_2$ between 40–50 mmHg. Blood should be available in the room as bleeding is a possibility.

6.5 Common atrioventricular canal (CAVC)

It is also called endocardial cushion defect and results from failure of endocardial cushions to fuse with lower part of atrial and upper portion of ventricular septum. In addition, atrioventricular valves will also be abnormal. There are three different types of CAVC exist.

1. Partial atrioventricular canal or Ostium primum defect
    a. Usually interatrial communication and cleft mitral valve
    b. Two separate AV valves
2. Transitional atrioventricular canal defect
    a. Ostium primum plus
    b. Partially separated AV valves
    c. Small to moderate VSD partially closed by chordate attachment
3. Complete atrioventricular canal defect
    a. Large non restrictive ostium primum
    b. Large VSD
    c. Large common single atrioventricular valve

Left to right shunting and regurgitation leads to volume loading of both atria and both ventricles. Patient will develops pulmonary congestion and pulmonary hypertension. Surgery usually performed at the age of 2-5 years and in some cases earlier.

Anaesthetic management

Management depends on severity of pulmonary hypertension and degree of left to right shunting. FiO2 and ventilation is manipulated along with use of NO and analgesics to reduce pulmonary hypertension. Ionotropic support will be required after bypass. TEE will be useful in detecting residual defects and ventricular function. LA line along with other invasive lines will help in deciding about escalation in ionotropes.

6.6 Anomalous pulmonary venous connection

Two types of abnormal communication are seen. Both of these defects may be associated with other cardiac lesions like ASD, VSD and PDA.

1. Partial anomalous pulmonary venous connection
    a. At least one pulmonary vein is connected to right atrium either directly or indirectly. Most common is right upper pulmonary vein opening in the superior vena cava. These patients may remain asymptomatic for long time.

2. Total anomalous pulmonary venous return (TAPVR)
    a. All four pulmonary veins opens in the right atrium.
    b. Four types of TAPVR exist
        i. Supra cardiac
            - Pulmonary veins converge and drains into a vertical vein which then drains into right atrium via innominate V or SVC
        ii. Cardiac
            - Common pulmonary confluence drains into coronary sinus
        iii. Infra cardiac or infra diaphragmatic
            - A common confluence of pulmonary vein passes through diaphragm and drains in the portal system which then drains into inferior vena cava.
        iv. Mixed
            - Pulmonary veins drains at two or more levels.

Pathophysiology of TAPVR depends on obstructed or non obstructed pulmonary venous return. Obstruction will leads to pulmonary venous hypertension and higher back pressures.

Anaesthetic considerations

PAPVR is associated with higher pulmonary blood flow, so main aim would be to reduce pulmonary blood flow. Patients with obstructed TAPVR are sicker and will need higher PaO2, ionotropic support and repeated blood gases to control acidosis. Post bypass period require high PaO2, hyperventilation, ionotropic support, good sedation, paralysis and NO. Intraoperative TEE is usually not done to avoid further obstruction of pulmonary veins but TTE and epicardial echo can be performed to look at venous return in the left atrium.

6.7 Transposition of the great vessels

Transposition is a common CHD which is associated with high mortality without intervention. Atrial septostomy is usually performed in the catheterization laboratory to stabilize the patient before surgery. PgE1 should be continued before bypass to keep the duct opens. Coronary artery[21] should be preoperatively assessed as abnormal location of coronaries creates surgical difficulties.

Anaesthetic considerations

Anaesthetic goal is to avoid reduction in cardiac output and systemic vascular resistance while keeping the PVR lower relative to SVR. Increased pulmonary blood flow due to reduced PVR will leads to increased mixing of blood and better saturation. Pulmonary resistance can be reduced by following measures:

1. Inhaled nitric oxide (NO)
2. Nebulized PGI2
3. Sildenafil (Oral and preferably intravenous)
4. Ventilatory interventions
    a. Increased FiO2
    b. Reduced carbon dioxide

c. Alkalotic PH
5. At the same time avoid hypoxia, hypercarbia, acidosis, hypothermia, high and low tidal volume, high PEEP and hypoglycaemia in neonates.

7. Postoperative pain management

High dose opioids are given during paediatric congenital heart surgery and analgesic effect continues in postoperative period. Good intraoperative and postoperative analgesia is associated with improved surgical outcome[22]. Morphine in the doses of 25 microgram/kg/hr will provide adequate analgesia and moderate sedation during postoperative period while additional sedatives are needed in intubated patients. Larger doses are needed in infants and young children basically due to high clearance.
Fentanyl at the doses of 1-5 microgram/kg/hr can be given instead to provide continuous analgesia but associated with less sedation then morphine. Non opioid analgesics like acetaminophen and ketamine can also be used as an adjunct to opioid analgesia. Ketamine is given intravenously as a bolus or in the form of continuous infusion at the rate of 10 – 45 microgram/kg/min.

8. Anaesthesia for catheterization laboratory procedures

Diagnostic and interventional cardiology plays a major in the management of congenital heart patients. General anaesthesia for these procedures is associated with low risk of morbidity and mortality[23]. Some of the challenges faced by anaesthesiologist in cath lab include

1. Usually located far away from operating rooms
2. Not equipped with recovery room
3. Transfer of critically ill patient from intensive care to cath lab or vice versa can create several problems
4. Rooms are usually undersized
5. Not properly illuminated
6. Access to patients airway is difficult
7. Monitoring interference with cath lab equipment
8. Contrast material use and its complications like contrast induced nephrotoxicity and vasovagal reaction
9. Radiation exposure
   a. Always use shielding devices like gown, glasses and thyroid collar
   b. Keep a distance from radiation source
   c. Minimize exposure time
   d. Different resident and consultant should rotate rather than assigning one person for cath procedures

Catheterization procedures should only be performed in centers where facilities for paediatric heart surgery are available. Catheterization procedures can be performed under local, monitored anaesthesia care (MAC) and general anaesthesia. There are several difficulties which make these procedures lengthy and complicated. Difficulties during procedure vary from intravenous access by anaesthesiologist to arterial and venous access by cardiologist. Necessary equipment for intubation and drugs for resuscitation should be

available as cardiac arrest in these patients is not uncommon[24] when sedation is given for the procedure.

Although cardiologists do give sedation for certain procedures but another person with ability to resuscitate the patient should be available in cath lab. Laryngeal mask airway (LMA) is well tolerated by most of the patients but those patients who can develop airway obstruction (Down's syndrome) during procedure should be intubated before the start of procedure. Intubation should also be done on those patients who need TEE. Anaesthesiologist has to be very careful during TEE manipulation. High doses of analgesia are not required and only local anaesthesia infiltration at access site is sufficient.

9. Complications

Expected complications during the procedure include

1. Arrhythmias
   a. Mechanical reasons, electrolyte disturbance and hypercarbia
2. Brachial plexus neuropathy due to stretching of nerve plexus during positioning
3. Hypothermia
4. Vascular damage at access site
5. Bleeding
6. Congestive heart failure
7. Tamponade

Use of ionotropes and vasopressors should be intentionally reduced, when anaesthesia is given for electrophysiological studies to minimize arrhythmias.

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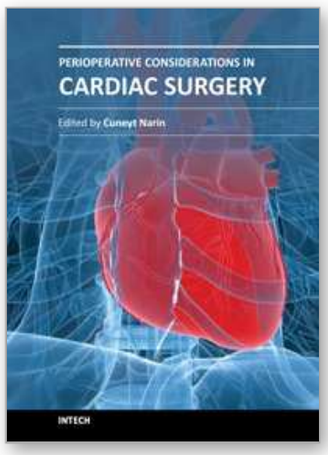

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This book considers mainly the current perioperative care, as well as progresses in new cardiac surgery technologies. Perioperative strategies and new technologies in the field of cardiac surgery will continue to contribute to improvements in postoperative outcomes and enable the cardiac surgical society to optimize surgical procedures. This book should prove to be a useful reference for trainees, senior surgeons and nurses in cardiac surgery, as well as anesthesiologists, perfusionists, and all the related health care workers who are involved in taking care of patients with heart disease which require surgical therapy. I hope these internationally cumulative and diligent efforts will provide patients undergoing cardiac surgery with meticulous perioperative care methods.

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