Review Article

Congenital heart diseases and anaesthesia

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ABSTRACT

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.

Key words: Anaesthetic management, cardiac catheterisation, congenital heart disease, Fontan physiology, left-to-right shunt, non-cardiac surgery, pulmonary hypertension

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.[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.[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 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.[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.[4] For uneventful anaesthesia in these patients, we need to understand the physiology and recognise the associated risks.[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

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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[7] [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[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 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 1st or 2nd year of life and then a Fontan procedure – both inferior and superior vena cava connected to PA. The

Table 1: Congenital heart disease broad classification

Acyanotic CHD	Cyanotic CHD	Shunt	No shunt
VSD	TOF	Increased Pulmonary Blood flow (Lt to Rt)	Obstruction to blood progression
ASD	TGA	Septal defects without pulmonary obstruction	AS, PS, CoA
PDA	TAPVR	Acyanotic group tIII Eisenmengerisation	Regurgitant lesion:
AVSD		Decreased Pulmonary Blood flow (Rt to Lt)	Mitral valve with AVSD, ASD
AP window		Septal defects with pulmonary obstruction	
		Cyanotic Group	
PS	Tricuspid atresia	Uncommon: Each <1% of CHD, pulmonary atresia	
AS		Ebstein's anomaly	
CoA	Truncus arteriosus		

VSD – Ventricular septal defect; ASD – Atrial septal defect; PDA – Patent ductus arteriosus; TGA – Transposition of the great arteries; TAPVR – Total anomalous pulmonary venous return; AVSD – Atrioventricular septal defect; AP – Aortopulmonary; CHD – Congenital heart disease; PS – Pulmonary stenosis; AS – Aortic stenosis; CoA – Coarctation of the aorta; TOF – Tetralogy of Fallot

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.[10] Children and adults with heart disease are at increased risk of mortality and morbidity when undergoing non-cardiac surgery.[4,9] Risk associated with an individual patient is based on several criteria [Table 2].[4,5,8]

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.[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.[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].

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

Table 2: Risk classification of children with congenital heart disease undergoing non-cardiac surgery

Risk	Physiology	Complexity lesions	Type of surgery	Age (years)	Timing/ASA	Pre-operative hospital stay
High For Adults PHT Cyanotic disease EF <35% LV outlet Obstruction	Physiologically poorly compensated and/ or presence of major complication like Cardiac failure Pulmonary hypertension Arrhythmia Cyanosis	Single ventricle or balanced circulation physiology Cardiomyopathy AS	Major Intraperitoneal Intrathoracic Major blood loss and anticipated transfusions	<2	Emergency surgery ASA IV-V	Pre-operative hospital stay >10 days
Intermediate For Adults Prosthetic Valve or conduit Moderate LV outlet obstruction	Physiologically normal and well compensated	Simple lesion	Major Intraperitoneal Intrathoracic Major blood loss and anticipated transfusions	<2	Emergency surgery ASA IV-V	Pre-operative hospital stay >10 days
Low	Physiologically normal and well compensated	Simple lesion	Minor or body surface surgery	>2	Elective ASA I-III	Pre-operative hospital stay <10 days

ASA – American of Anesthesiologists; AS – Aortic stenosis; PHT – Pulmonary hypertension; EF – Ejection fraction; LV – Left ventricular

Table 3: Pre-operative assessment and investigations

Class of surgery	Blood investigations	Physical examination	Specific investigation	Pre-operative medications
Catheterization without intervention For intervention in high and moderate risk	Hb, coagulation profile Renal function test Liver function test Blood group and save packed cells (one adult unit) for any interventional procedure	BP in all extremities (for CoA or PDA) Baseline oxygen saturation in all limbs and effect on oxygen saturation due to exercise. Oedema, murmur, crackles	Chest X-ray Echo within 1 month is enough or one day prior to procedure Review CT MRI	Continue all pre-operative medications like digoxin Propranolol ACE inhibitors Bosentan Sildenafil
Non-cardiac surgery Major, semi-major surgery High to intermediate Risk patients	All blood investigations Blood group and save a cross-match of packed cells	Baseline oxygen saturation in all limbs and effect on oxygen saturation due to exercise. Oedema, murmur, crackles	Chest X ray Recent echo	Continue all pre-operative medications such as digoxin Propranolol Bosentan, sildenafil ACE inhibitors may be stopped
Grown up cardiac patient for surgery	All blood investigations Blood group and antibody screen should be performed for all and a cross-match of packed cells	Baseline oxygen saturation in all limbs Oedema, murmur, crackles, reduced exercise tolerance	Chest X ray The most recent echocardiography and MRI reports should be reviewed	Continue all PHT reducing medication like bosentan, sildenafil ACE inhibitors should be stopped unless short duration procedure no major fluid shift

PDA – Patent ductus arteriosus; CoA – Coarctation of the aorta; MRI – Magnetic resonance imaging; CT – Computed tomography; ACE – Angiotensin-converting enzyme; PHT – Pulmonary hypertension; Hb – Haemoglobin; BP – Blood pressure

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.[13]

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

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[8] can present as follows:

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

Table 4: Post-cardiac surgery - long-term problems

Disease	Procedure	Long-term sequelae
ASD	Transcatheter device closure	Residual shunt, device fracture, malposition, septal aneurysm
Aortic coarctation	Surgical correction or Balloon dilatation	Systolic hypertension, residual gradient, left arm pressure low due to sub-clavian flap. Aneurysm formation
TGA	Arterial switch operation	Supravalvular AS PS, aortic insufficiency, coronary artery stenosis, ventricular dysfunction
	Senning/mustard	Arrhythmia, right ventricular (systemic) dysfunction, Baffle leak, obstruction
	Rastelli procedure	Residual VSD, ventricular dysfunction, outflow tract obstruction, aortic and pulmonary valve dysfunction
TOF	BT shunt VSD closure and transannular patch, unicuspid valve	Ipsilateral arm BP and SpO_2 inaccurate, RBBB, AV block, pulmonary insufficiency, RV dysfunction, arrhythmias. Sudden death
Single ventricle disease	Fontan operation	Preload dependence, ventricular dysfunction, elevated PVR-right heart failure, cyanosis hepatomegaly, protein losing enteropathy, exercise intolerance
Heart failure	LVAD Heart transplant	Anti-coagulation problems Ventricular dysfunction: Rejection, coronary artery disease, recurrent infections, malignancy, complications of immunosuppression. Glucose intolerance

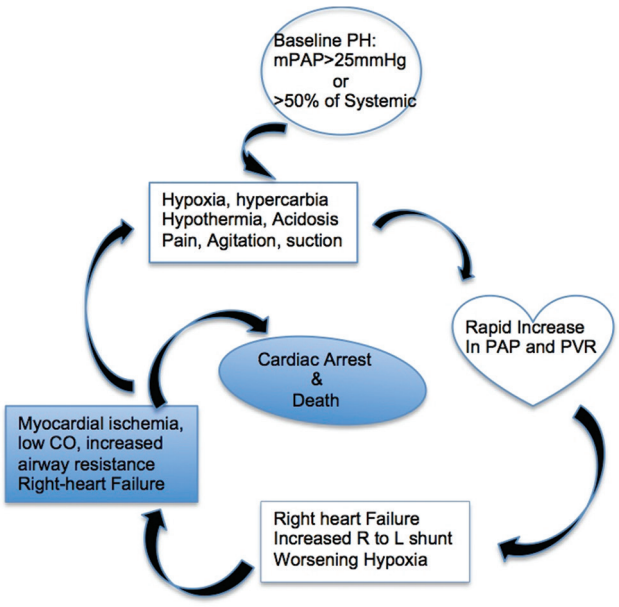
ASD – Atrial septal defect; TGA – Transposition of great arteries; TOF – Tetralogy of Fallot; BT – Blalock–Taussig; VSD – Ventricular septal defect; LVAD – Left ventricular assist device; BP – Blood pressure; AS – Aortic stenosis; PS – Pulmonary stenosis; RBBB – Right bundle branch block; AV – Atrioventricular; RV – Right ventricle; PVR – Pulmonary vascular resistance

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.[11] Avoiding sympathetic stimulation and use of nitric oxide can be lifesaving in these patients [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.



Vicious Cycle of Pulmonary Hypertension leading to cardiac arrest
mPAP: Mean Pulmonary Artery Pressure, PVR : Pulmonary Vascular resistance, CO : Cardiac Output

Figure 1: Vicious cycle of pulmonary hypertension

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):[8]

1. Patients with prosthetic cardiac valves
2. Patients with prior infective endocarditis
3. Patients with unrepaired or palliated cyanotic CHD including shunts and conduits
4. Patients with CHD repair with prosthetic material or device placed by surgery or catheter intervention during first 6 months after placement
5. 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.[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.[16]

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.[19]

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 μg/kg) are required to blunt haemodynamic

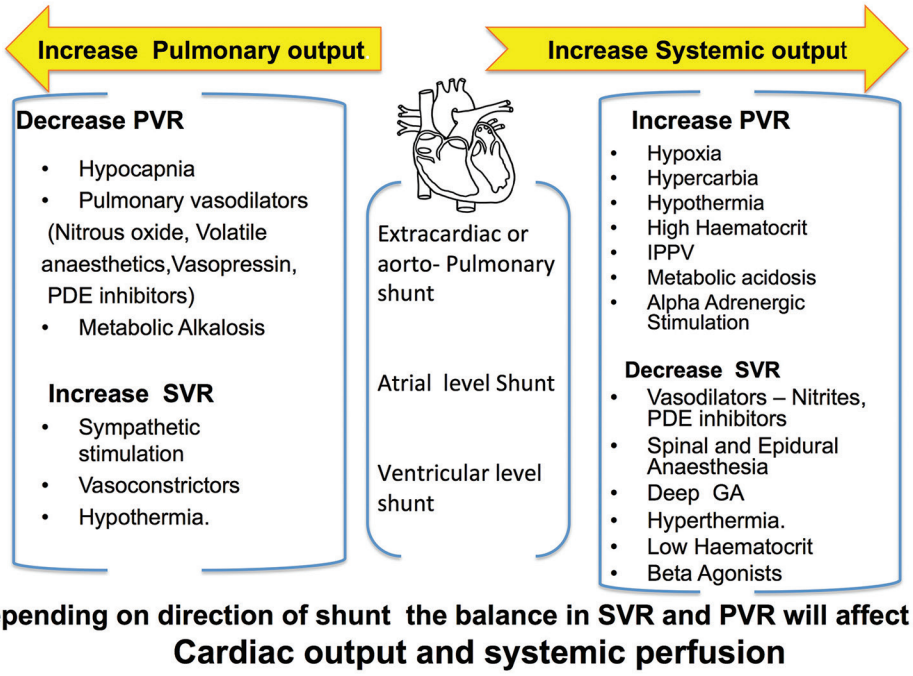


Figure 2: Flow distribution dynamics. (PVR: Pulmonary vascular resistance; SVR: Systemic vascular resistance)

Table 5: Anaesthesia management (Note: Deairing of intravenous line is very important!)

Congenital heart disease: Surgical group	Physiology	Potential issues	Choice of agents	Specific precautions in anaesthesia management
Diagnostic and therapeutic cardiac catheterization (e.g. BAS, ASD device closure, measurement of PVR, balloon valvotomy-pulmonary or aortic, CT angio studies for MAPCAs)	Uncorrected defects shunting right to left or left-to-right Balance or single ventricular physiology May be associated with other congenital defects (like airway)	Desaturation PHT During oximetry use of low oxygen makes it difficult while resuscitation Arrhythmia due to catheters manipulation chances of blocks	Ketamine and sevoflurane to adjust SVR Dexmedetomidine for sedation anaesthesia[11] Remifentanil for short procedure and clear headed recovery	Avoid excessive tachycardia and bradycardia For TOF, maintain SVR Avoid inotropes to reduce chances of infundibular spasms[17] Be aware of cyanotic spell which may need vasopressors such as phenylephrine and morphine for management
Uncorrected cardiac cases for non-cardiac cases (e.g. infant with VSD, ASD for cleft lip/palate repair, circumcision, scrotal swelling, ToF child with brain abscess)	Uncorrected defects Shunting Balance or single ventricular physiology May be associated with other congenital defects	Desaturation PHT Arrhythmias	SVR fall needs to be avoided: etomidate for induction If intubation needed, then cisatracurium-atracurium or atracurium as choice of relaxant Fentanyl or remifentanil for pain relief or dexmedetomidine Spinal anaesthesia can be used in few type of surgeries. Evidence shows that few types of CHD patient can tolerate spinal anaesthesia[18]	Avoid excessive tachycardia and bradycardia For TOF, maintain SVR Avoid inotropes to reduce chances of infundibular spasms For surgeries with major fluid shift, invasive lines are a must For CVC line to avoid arrhythmias, length can be adjusted by formulae 0.07 × height in cm + 1.7 cm May need inotropic support in perioperative period in the form of dopamine, dobutamine, adrenaline infusion
Corrected cardiac condition and anaesthesia (e.g. repaired ASD, VSD, ToF, single ventricle	Normal physiology if complete correction Single ventricular physiology for Fontan	Minimal issues with normal physiology For single ventricle need to maintain passive pulmonary flow with spontaneous ventilation	No restriction for the use of anaesthetic agents	For Fontan procedure, avoid fluid overload but maintain preload!! If using IPPV avoid high Inspiratory pressures and prolonged inspiratory time Keep ventilation near FRC and maintain normocapnoea

BAS – Balloon atrial septostomy; ASD – Atrial septal defect; PVR – Pulmonary vascular resistance; CT – Computed tomography; MAPCAs – Major aortopulmonary collateral arteries; VSD – Ventricular septal defect; TOF – Tetralogy of Fallot; PHT – Pulmonary hypertension; SVR – Systemic vascular resistance; CHD – Congenital heart diseases; CVC – Central venous catheter; IPPV – Intermittent positive pressure ventilation; FRC – Functional Residula capacity

Table 6: Anaesthetic drug effects and doses

Anaesthetic drug	SVR	PVR	Dose
Potent volatile agents	↓	↓	0.5-1 MAC
Nitrous oxide	-	↑	
Opioids			
Fentanyl	↓		1-2 μg/kg 0.5-2 μg/kg/h
Morphine	↓		0.05-0.2 mg/kg 0.02-0.2 mg/kg/h
Midazolam	-	-	0.5 mg/kg PO 0.1 mg/kg IV 0.02-0.3 mg/kg/h
Ketamine (with glycopyrrolate)	↑	No effect (if CO_2 normal)	3-4 mg/kg PO 1-2 mg/kg IV 5-20 μg/kg/min
Propofol	↓	-	2-3 mg/kg 100-300 μg/kg/min
Dexmedetomidine	↑-↓	↓	1 μg/kg (over 10 min) 0.2-1 μg/kg/h

IV – Intravenous; SVR – Systemic vascular resistance; PVR – Pulmonary vascular resistance; MAC – Minimum Alveolar Concentration ; ↓ – lowers; ↑ – increases

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.[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).[19] Respiratory events are more common in infants and intubated patients.[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[20,21] [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_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.[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 $PaCO_2$ 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.

Table 7: Complications after cathlab-related procedure

Type of complication	Example
Haemodynamic	Cardiac arrest, hypoxia, acidosis
Arrhythmia	Bradycardia, SVT, VT, VF, AV blocks, junctional rhythm
Vascular	Thrombosis, arterial, venous tears, perforations
Bleeding	Haematomas at catheter site or retroperitoneal
Catheter manipulation related	Cardiac perforation, tamponade, air embolism, wire break and knot
Device related	Device embolisation, balloon rupture
Other	Drug interactions, infection, cerebral events

SVT – Supraventricular tachycardias; VT – Ventricular tachycardias; VF – Ventricular fibrillation; AV – Atrioventricular

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