11 October 2019
No. 19

Considerations in Congenital Heart Disease for Non-Cardiac Surgery

Dr Arusha Maharaj

Moderator: Dr Avintha Ramkisson

UNIVERSITY OF
KWAZULU-NATAL

INYUVESI
YAKWAZULU-NATALI

School of Clinical Medicine
Discipline of Anaesthesiology and Critical Care

CONTENTS

CONSIDERATIONS IN CONGENITAL HEART DISEASE FOR NON-CARDIAC SURGERY

Introduction

Congenital heart disease (CHD) is among the commonest birth defects, with approximately 1 in 125 live births per annum (1) and accounting for 28% of all major congenital abnormalities (2). Limited healthcare access and therefore diagnostic facilities in many parts of the world, may account for the differences in reported birth prevalence between high and low income countries.

The cause of most CHD is unknown but can be related to genetic defects, teratogens (maternal exposure to alcohol, drugs) or maternal disease(3). South Africa has an estimated 11000 children with CHD born annually(4). With appropriate care and advances in medical care (1, 4) 85% to 90 % of these children are expected to survive to adulthood and therefore prognosis is excellent. However, one has to account for the challenges faced by developing countries in providing this care. Of the children with CHD who rely on public health services in South Africa, less than 25 % of them receive the care they need (4).

Apart from these children requiring surgery (most often in the first year of life), due to extra cardiac anomalies such as tracheo-oesophageal fistula, anorectal malformations, cleft lip and palate, renal and skeletal pathologies (5), they are also subject to the same childhood illnesses as healthy children and may present to the local hospital requiring emergency and elective surgery(6). Significant advances have been made in the diagnosis and treatment of children with CHD over the past decades. This means an increase in life expectancy for this group of patients who will increasingly seek medical attention for other illnesses as well as undergo non- cardiac surgeries(7).

The Paediatric perioperative cardiac arrest registry (POCA), collected data on 373 anaesthesia related cardiac arrests in children, between 1994 to 2005 , 34% (n=127)of whom had congenital or acquired heart disease. This retrospective study was not powered to compare the cardiac arrests between acquired versus congenital heart disease however, of the cardiac arrests that occurred in the heart disease group, structural CHD accounted for 88 of the 127 cardiac arrests. The lesions with the highest rates of cardiac arrest were single ventricle (19%), left to right shunt lesions (18%) and obstructive lesions (16%). Furthermore, the majority of cardiac arrests in children with heart disease were reported most frequently from the general operating rooms (54%) as compared to 46% of cardiac arrests that occurred in the cardiac operating rooms, cardiac catherisation laboratories and other imaging suites(8). A more recent review compared (7) the post-operative outcomes of children with and without CHD undergoing non-cardiac surgery. The incidence of postoperative mortality was significantly higher in children with moderate (3.9%) and severe (8.2) CHD compared with their controls, 1.7 % and 1.2 % respectively. No difference was observed between children with minor CHD and their matched controls.

The complexity of cardiac lesions, the variety of non -cardiac surgery and the pace of change in the field of cardiac surgery makes a one size fits all approach to evidence based perioperative management impossible(6). Practitioners who care for patients with CHD must address a number of challenges including identifying the best location for procedures (e.g. a children's hospital) identifying a suitably qualified team (cardiac anaesthetists, surgeons), improving expertise of the non-cardiac sub-specialities(paediatricians , nephrologist etc.)(7).

The recognition of high risk patients, multidisciplinary decision making as well as an understanding of the anatomy, pathophysiology of the CHD and palliative procedures are critical for optimising outcomes in this patient population during the peri-anaesthetic period.

Foetal and transitional Circulation:

Any discussion of congenital heart lesions would not be complete without a revision of foetal and transitional circulation.

The cardiopulmonary circulation in utero is a parallel circulation as compared to the series circulation of the neonate.

The parallel circulation is made possible by 3 shunts:
Ductus venosus,
Foramen ovale and
Ductus arteriosus.

Oxygenated blood from the placenta returns to foetal circulation (O_2 saturation 80%) via the umbilical vein. 30% of this blood passes via the ductus venosus to the inferior vena cava (IVC). Due to the porto-caval gradient this blood accelerates and pushes open the second shunt: foramen ovale, to enter the left atrium (LA) from the right atrium (RA). This preferential streaming allows blood (O_2 saturation of 65%) to pass to the left of the heart , then to the aorta to supply mostly the coronary, subclavian and carotid arteries(9).

Poorly oxygenated blood from the upper part of the body returns via the superior vena cava (SVC) and is preferentially directed to the right ventricle (RV). This blood (O_2 saturation of 55%) is then pumped to the pulmonary artery. Due to the high pulmonary vascular resistance (PVR), 95% of RV blood is shunted via the ductus arteriosus, into the descending aorta and back to the placenta and lower body(10).

Note that 30% of blood from umbilical vein is directed to the RA and the rest is redirected to the liver. This suggests that the liver is a developmental priority for well oxygenated blood(9).

The ductus arteriosus is a muscular blood vessel which connects the pulmonary arterial trunk to the aorta, and its patency is under the influence of circulating prostaglandins and nitric oxide. Foetal pulmonary arteries are thick walled and high vasomotor tone therefore having high PVR. Less than 20% of foetal cardiac output (CO) does go to the lung in order to support growth and metabolism and as the pulmonary vascular bed enlarges the percentage of cardiac output via the lungs increases(9).

Closure of Shunts:

Ductus venosus: Closes 1 to 3 weeks post-delivery, and later in premature infants.

Foramen ovale: undergoes functional closure when there is reversal of the PVR to SVR pressures at deliver. It however remains probe patent in the majority of infants and up to 30% of adults.

Ductus arteriosus: contracts in response to increased O_2 tension: and is functionally closed 10 to 15 hours post-delivery. Anatomical closure is by fibrosis at 4 to 6 weeks post-delivery(9).

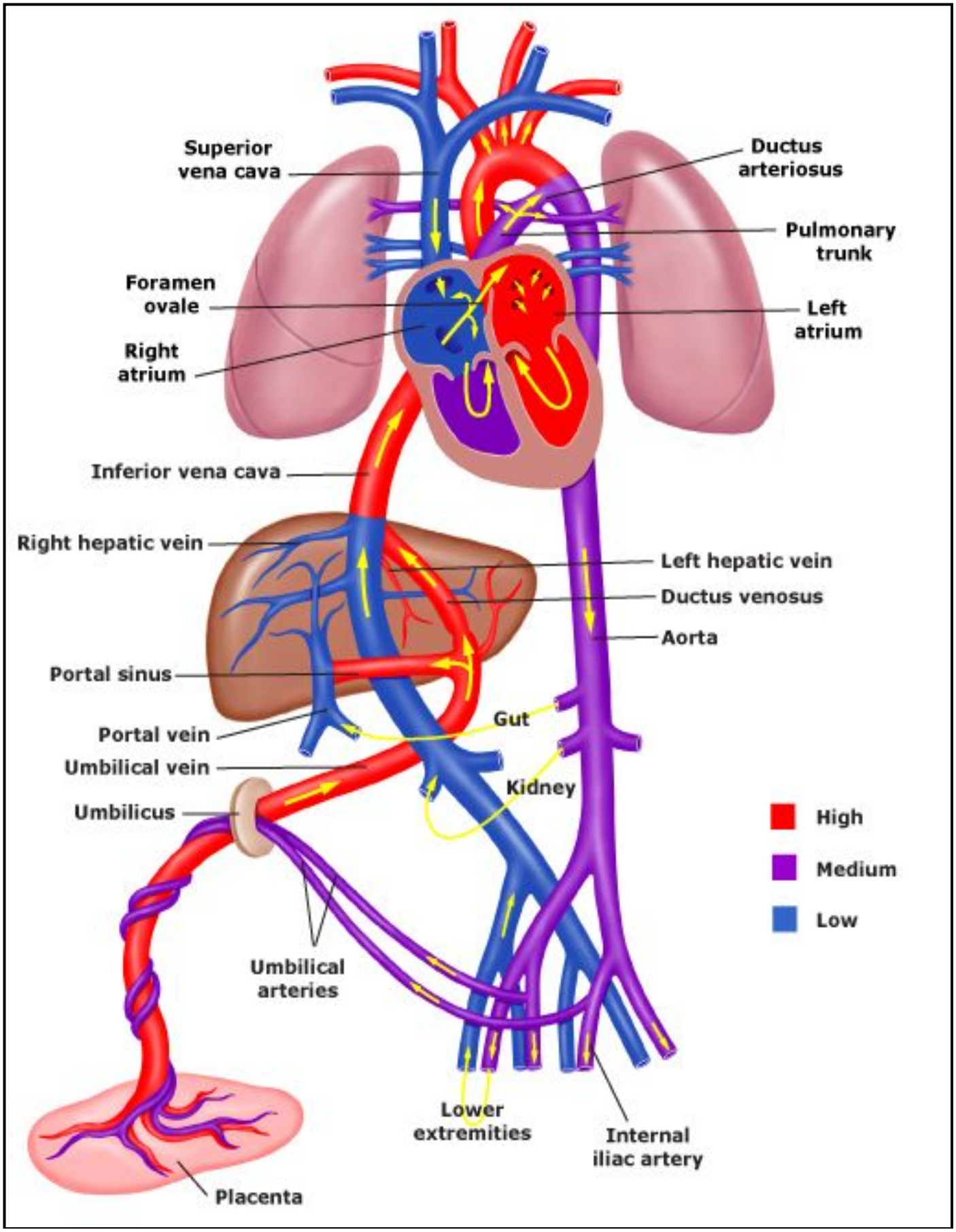


Figure 1: Foetal circulation (11)

Classification of CHD:

In a broad context CHD can be classified as structural defects and conduction defects.

The definition of structural congenital heart disease is “a gross structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance” (2) and present from birth.

Ventricular septal defects (VSD) are the commonest structural defects in all populations although incidence of the other defects varies across populations(3).

Conduction defects as a form of CHD is due to the presence of abnormal conduction pathways which lead to the development of arrhythmias. Common congenital conduction defects are:

a. supraventricular tachycardias (leading to Wolf- Parkinson White Syndrome characterised by a short PR interval on ECG)
b. Ventricular tachycardias (often associated with cardiomyopathy and Torsade’s de Pointes)
c. Congenital bradycardia (<90 beats per min)(3)

However for the purposes of this review we will focus on structural CHD.

The following classification (based on anatomical defect) has been taken from the review article by Chowdhury(12).

1.	Left to right shunts
	Atrial level: ASD, TAPVC
	Ventricular level: VSD
	Great artery level: PDA, AP window,
	Truncus arteriosus
	Coronary level: ALCAPA, coronary fistula
2.	Right to left shunts
	TOF physiology
	TGA physiology
3.	Left heart obstructive lesions
	Obstructed veins
	Mitral stenosis
	Aortic stenosis
	Coarctation
	Interrupted aortic arch
	Hypoplastic left heart syndrome
4.	Right heart obstructive lesion
	Pulmonary stenosis / atresia
	Tricuspid stenosis
	Hypoplastic right heart
5.	Single ventricle
6.	Others
	Vascular rings
	Venous anomalies
	Arteriovenous fistulae

Table 1: Classification of CHD based on anatomical defect.

Structural defects can also be classified in a simpler, more practical manner as cyanotic or acyanotic CHD and further subdivided on the basis of the pulmonary and systemic blood flow.

Acyanotic	**Cyanotic**
VSD	ToF
ASD	TGA
PDA	TAPVC
AVSD	Tricuspid atresia
PS	Truncus arteriosus(TA)
AS	Pulmonary atresia
Co-arctation of the Aorta	Ebstein's anomaly

Table 2: Classification of acyanotic and cyanotic CHD (13).

Lesions that cause excessive pulmonary blood flow: VSD, ASD, and PDA.
Lesions that cause inadequate pulmonary blood flow: ToF, Pulmonary atresia.
Lesions that cause inadequate systemic blood flow: CoA. (13)

Critical CHD refers to defects that require surgical correction in the first year of life(14).

Statistics regarding incidence specific to South Africa are difficult to find and hence UK (3) and worldwide prevalence (2) are quoted.

Lesion	**United Kingdom**	**Worldwide**
VSD	32%	34%
PDA	12%	10%
PS	8%	8%
CoA	6%	5%
ASD	6%	13%
ToF	6%	5%
AS	5%	4%
TGA	5%	5%
HLHS	3%	
HRHS	2%	
AVSD	2%	
TA	1%	

Table 3: Prevalence of congenital heart disease.

Principles of Pathophysiology in CHD:

Aberrations in cardiac anatomy are complex and diverse.

Normal (series) versus balanced (parallel) circulation:

In a normal circulation the pulmonary and systemic circulations are in series. This means that deoxygenated blood to right heart passes to the lungs and oxygenated blood returns from the lungs to the left heart to be pumped to the systemic circulation. Some forms of CHD can have a normal / series circulation with one or more defects that lead to mixing and shunts. The circulation

can become a balanced/ parallel circulation in the face of a large unrestrictive defect or with a change in the relative resistances of the pulmonary and systemic circulations. For illustrative purposes: a balanced/parallel circulation can be created during anaesthesia while using high concentrations of inspired oxygen. This creates a decrease in PVR allowing a left to right shunt to worsen. Hence the term balanced circulation: as blood flow to the lungs and systemic circulation is a balance between PVR and SVR(6).

a. Left to right Shunting:

   These are the most common lesions and are classified as acyanotic. The size of the lesion and the pressure gradient between the higher left sided systemic pressure and lower right sided pulmonary pressure determine blood flow (Q) across the shunt. With an increase in flow across the shunt there is an increase in pulmonary circulation. Therefore venous return to the LV increases with volume overload and decreased systemic flow (12). The heart has to augment its cardiac output to compensate for the proportion of flow that recirculates to the pulmonary system(14). Signs of heart failure will become evident when pulmonary flow (Qp) exceeds systemic flow (Qs) by the 3:1(14). During foetal life even a large defect will not cause major physiological effect due to the high PVR restricting pulmonary flow. Transition to extra-uterine circulation (decrease PVR and increased SVR) will allow manifestation of left to right shunts in the form of congestive cardiac failure. The physiological nadir in haemoglobin occurring at 3 months also exaggerates features of heart failure(12).

b. Right to left shunting:

   These are classified as cyanotic lesions, since deoxygenated blood is distributed to the systemic circulation. Most commonly these shunts will develop from a long standing uncorrected left to right shunt. The increased pulmonary blood flow causes progressive structural changes in the pulmonary vascular bed and therefore ultimately pulmonary hypertension. The increased right sided pressures eventually exceed the left side pressure (supra- systemic) and the shunt becomes a right to left shunt. Often referred to Eisenmenger's syndrome(14). There are however, lesions that begin as right to left shunts such as ToF. These are discussed later.

c. Obstructive lesions:

   Many of the patients may initially be asymptomatic i.e. if there is no critical obstruction, e.g. aortic stenosis, pulmonary stenosis. To compensate for the fixed outflow obstruction the ventricle becomes hypertrophied, leading to decreased compliance and diastolic dysfunction. Symptoms may become evident when features of diastolic dysfunction set in(14).

d. The single ventricle:

   There are several variations (e.g. Tricuspid Atresia, hypoplastic left heart syndrome, etc.) Depending on the lesion, there will be complete mixing of oxygenated and deoxygenated blood. Also the quantity of pulmonary blood flow will be determined by the degree of pulmonary stenosis if present. Without pulmonary stenosis, Qp is far greater than Qs as PVR is much lower than SVR. This leads to increased left to right shunting and congestive cardiac failure(12).

   With pulmonary stenosis, blood flow to the lungs is restricted and therefore a patent ductus arteriosus is required to maintain adequate pulmonary flow. Blood will flow down the

pressure gradient via the ductus arteriosus to the PA's, or with a lower degree of RVOT obstruction a balance circulation may exist where Qp: Qs is 1:1 and 2:1(12).

Important to remember is that in any of these, the single ventricle is pumping the full cardiac output (CO). CO = Qp + Qs. Thus the ventricle is volume overloaded. Those variations with right ventricular outflow tract (RVOT) obstruction will require a Blalock-Taussig's (B-T) shunt as palliation for the decreased pulmonary flow and the volume overloaded ventricle(12). This and other palliative procedures are summarised later.

Three types of patients with CHD:

In practice one can expect to encounter one of three types of CHD patients:

a. Patient with an uncorrected cardiac lesion
b. Patients who have had palliative surgery e.g. ToF with a B-T shunt
c. Patients with corrected CHD lesion with or without residual defects. (8, 13, 14)

Peri-operative considerations:

As dictated by professional responsibility the preoperative evaluation must include a detailed account of the anatomic cardiac lesion, the altered physiology and the implications there-of. The following is a guide to the preoperative evaluation(1, 13).

a. What is the anatomical change associated with the cardiac defect and any palliative procedures? (this can be done via a review of all relevant investigations:(blood tests, radiography, echocardiology reports, catheterisation reports)
b. Direction and amount of shunt?
c. The extent of increase or decrease of pulmonary blood flow, if there is an increase in pulmonary blood flow is pulmonary hypertension present or if there is a decrease in blood flow, is there a shunt present?
d. What is the degree of hypoxia and polycythaemia?
e. Do any coagulation defects exist or is there evidence of thromboembolic phenomena?
f. Functional status of patient?
g. What associated pathological findings will influence management?
h. Review of drug history: anti failure therapy, anticoagulants, and anti-arrythmic agents. These agents have an impact on peri-operative management.
i. Other factors such as appropriate counselling for family.

When considering what associated pathological findings will influence management, one also has to question if time allows for optimisation and how to accomplish this optimisation.

Important associated pathological findings and or complications of congenital heart disease are the presence of:

a. Cardiac failure
b. Pulmonary hypertension
c. Arrhythmias
d. Cyanosis

a. Cardiac Failure:

A continually volume overloaded or pressure overloaded heart will lead to cardiac failure. Limited cardiac reserve is when the heart functions at near maximal capacity even at rest. These patients are at high risk of cardiac failure during anaesthesia and dictate the avoidance of agents like propofol which have a deleterious effect on cardiac output. Conversely ketamine has minimal effect(1) as plasma noradrenaline levels in cardiac failure are increased. Ketamine supports sympathetic tone and results in stable haemodynamics(8).

The decreased cardiac output prolongs inhalational and intravenous induction, so patience is required to avoid excessive drug administration. There is also the risk of poor coronary perfusion and for this adrenaline or phenylephrine should be available (1). Venous access may also be difficult (6).

b. Pulmonary hypertension (PHT):

This is defined as a mean pulmonary artery pressure (PAP) above 25 mmHg at rest and above 30 mmHg on exercise(6) in the presence of equal distribution of blood flow to all lung segments (14).

Echocardiographic features suggestive of PHT include a tricuspid regurgitation Doppler velocity of more than 2.5m/s or an estimated systolic pulmonary artery pressure that is more than 50% of systemic systolic arterial blood pressure(15). Pulmonary hypertension deserves special mention as these patients are at increased risk from anaesthesia and surgery. As seen in the review by Ramakrishna(16) adults with pulmonary hypertension have a postoperative morbidity and mortality of 7% in non-cardiac surgery.

Patients at risk are those with:

i) a left to right shunt (excessive pulmonary blood flow)
ii) prolonged pulmonary venous obstruction,
iii) high left atrial pressure.

Those with supra-systemic PAP are eight times more likely to have a major complication than those with sub-systemic PAP. Pulmonary hypertension reduces airway compliance and increases airway resistance, which leads to an increase in work of breathing. With respiratory tract infections having a greater impact and being more poorly tolerated (1).

PHT causes fixed structural changes in the vascular bed with reactive smooth vascular muscle, that is affected by several factors (1). Appropriate management strategies that will reduce PVR, improve RV function and decrease right to left shunting depend on understanding of the specific intra-cardiac anatomy and physiological consequences.

A rapid increase in PVR can lead to a pulmonary hypertensive crisis with or without right heart failure. This can be life threatening especially at the point where PVR begins to supercede systemic pressure. PVR increases in response to a variety of stimuli including; alveolar hypoxia, hypoxemia (PaO_2 <60mmHg), hypercarbia, metabolic acidosis and activation of the sympathetic nervous system by noxious stimuli and hypothermia(1, 15).

Symptoms and signs of a pulmonary hypertensive crisis include syncope, dyspnoea, cyanosis, pallor, bradycardia, right ventricular heave and bronchospasm(15).

TREATMENT	RATIONALE
Administer 100% O_2	Increases PaO_2 and decreases PVR
Hyperventilate to induce a respiratory alkalosis	PAP is directly related to $PaCO_2$
Correct a metabolic acidosis	PVR is directly related to H+ concentration
Administer pulmonary vasodilators	To decrease pulmonary vascular resistance, first choice inhaled nitric oxide
Support cardiac output	Adequate preload and inotropic support addresses right ventricular function
Attenuate noxious stimuli	E.g. Pain can increase PAP

Table 4: Treatment strategy for pulmonary hypertensive crisis.
Adapted from Freisen et al(15).

The use of pre-operative pulmonary vasodilator therapy may modify peri-operative morbidity, but does not appear to impact mortality(14).

c. Arrhythmias:

Ventricular ectopics (VE) are an ominous sign as 30% of these patients die suddenly. Factors known to decrease threshold for VE must be avoided for example hypercarbia, acidosis, hypoxia and large doses of local anaesthetic with adrenaline(1).

Some operations are associated with an increased risk of late onset arrhythmias e.g. those with extensive atrial sutures or ventriculotomy having the highest risk. Necrosis and progressive fibrosis extending into the conduction system are possibly the cause and these can be associated with sudden collapse and death(1).

The types of arrhythmias to be anticipated are:

- Atrial arrhythmias: commonly with repair of sinus venosus defects, atrial septal defects or atrial switch procedures and total cavo-pulmonary anastomosis.
- Sub-atrial arrhythmias: ventriculotomy or right ventricle to PA conduit can result in damage to AV node and Bundle of His.
  The risks of these are reduced but not eliminated by modern surgical techniques and earlier repair(1).

Some patients may exist with a primary malignant arrhythmia like prolongation of the QT interval, associated with Torsade de Pointes. Evidence shows that propofol and sevoflurane do not affect the QT interval while desflurane prolongs corrected QT interval (QTc) in normal children(1).

d. Cyanosis:

Cyanosis is a common feature of congenital heart disease, either in the unrepaired or palliated state. It is seen commonly but not exclusively in lesions with right to left shunt due to the decreased pulmonary blood flow(PBF) and effects all major organ systems(1). The haematological consequences of chronic cyanosis will be discussed in detail.

Chronic hypoxia increases erythropoietin production which will lead to increases in haemoglobin and hence haematocrit and blood viscosity(1). The increased haemoglobin allows more oxygen delivery without a sustained increase in cardiac output. Above a haematocrit of 65% complications can arise such as decreased O_2 delivery from red cell rigidity (especially with iron deficiency) and hyper-viscosity leading to cerebral vein and sinus thrombosis. The high risk groups for such complications are: children under 5 years,

dehydration, iron deficiency and fever. To minimize risk preoperative fasting should be kept to a minimum, and intravenous fluid therapy is indicated(1).

Laboratory tests of haemostasis can be abnormal in up to 20% of children with cyanosis (1). Defects are:

- prolonged prothrombin time and partial thromboplastin time,
- thrombocytopenia,
- platelet dysfunction
- hypo-fibroginemia
- accelerated fibrinolysis.

Aspirin is often used to avoid shunt thrombosis and this should be continued in the perioperative period, as the risk of thrombosis is higher than bleeding (1).

Risk Stratification for non-cardiac surgery:

Perioperative risk is most importantly determined by the perioperative physiological status and the complexity of the cardiac defect. Patient age, comorbidities and procedure related risk will further influence defect associated risk(14).

Physiologically well compensated patients can tolerate elective procedures at low risk, however those who are poorly compensated undergoing major or urgent operations are at high risk(17). With this in mind, the range of CHD and a variety of non-cardiac surgeries make risk stratification difficult (6).

The following table (Table 5) can offer a practical and structured approach to risk stratification by classifying children into high, intermediate and low risk groups.

High risk	Intermediate risk	Low risk
Physiologically poorly compensated and/or presence of major complications (a) Cardiac failure (b) Pulmonary hypertension (c) Arrhythmias (d) Cyanosis	Physiologically normal or well compensated	Physiologically normal or well compensated
Complex lesions (single-ventricle or balanced circulation physiology, cardiomyopathy, aortic stenosis)	Simple lesions	Simple lesions
Major surgery (intraperitoneal, intrathoracic, anticipated major blood loss requiring transfusion)	Major surgery (intraperitoneal, intrathoracic, anticipated major blood loss requiring transfusion)	Minor (or body surface) surgery
Under 2 yr old	Under 2 yr old	Over 2 yr old
Emergency surgery	Emergency surgery	Elective surgery
Preoperative hospital stay more than 10 days	Preoperative hospital stay more than 10 days	Preoperative hospital stay less than 10 days
ASA physical status IV or V	ASA physical status IV or V	ASA physical status I–III

Table 5: Approach for risk stratification(6).

Note the presence of long term sequelae of cardiac failure, pulmonary hypertension, arrhythmias and cyanosis is also considered as defining complex disease(6).

Risk stratification and classification should then be able to assist in the decisions regarding site of surgery and degree of postoperative care. White and Peyton(6) outline a suggested management based on risk and elective versus emergency surgery in Table 6.

	Elective	***Emergency***
High risk	Transfer to specialist centre	Consult the multidisciplinary team: Paediatric intensive care transport team, paediatric surgeons regarding feasibility of transfer. If transfer is not feasible get advice from cardiologist and paediatric cardiac anaesthetist regarding perioperative management. Transfer when stable enough.
Intermediate risk	Discuss with specialist centre and consider transfer	As above.
Low risk	Manage at local hospital	Manage at local hospital (can consult the specialist centre for concerns).

Table 6: Suggested required level of care based on risk stratification(6).

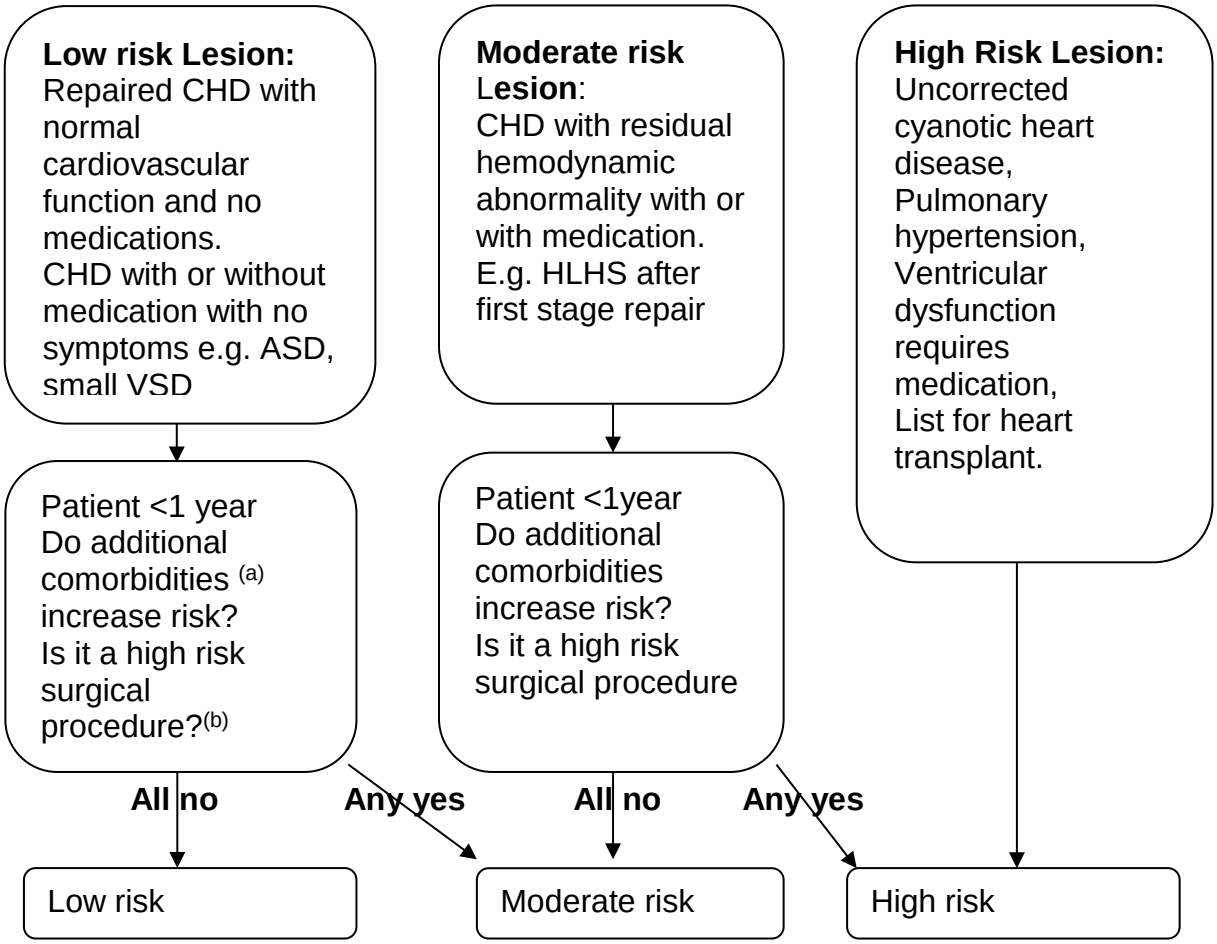
Transfer to a specialist centre is recommended when there is high likelihood of requirement for intensive care and on site cardiology support whereas, the transfer of a low risk patient creates unnecessary work for the specialist centre and places undue burden on the family (transport costs and lack of social support networks) (6).

American College of Surgeons (ACS), National Surgical Quality Improvement Programme (NSQIP), defines CHD more specifically as minor, major and severe. It also incorporates residual lesion and functional status.

Classification	Definition and Criteria
Minor CHD	• Cardiac condition with or without medication and maintenance (e.g., atrial septal defect, small-to-moderate ventricular septal defect with no symptoms) • Repair of congenital heart defect with normal cardiovascular function and no medication
Major CHD	• Repair of congenital heart defect with residual hemodynamic abnormality with or without medications (e.g., tetralogy of Fallot with wide open pulmonary insufficiency, hypoplastic left heart syndrome including stage 1 repair)
Severe CHD	• Uncorrected cyanotic heart disease • Patients with any documented pulmonary hypertension • Patients with ventricular dysfunction requiring medications • Listed for heart transplant

Table 7: ACS NSQIP classification of CHD based on residual lesion burden and functional status(7).

Using the above classification Pilkington et al devised an alternative perioperative flowsheet for risk assessment.



Note:
Co-morbidities: Acute or chronic kidney disease, insulin dependent diabetes, significant developmental delay, or other comorbid illnesses.
Types of surgery: Vascular, thoracic, open abdominal with large amount of fluid shift, intra-parenchymal neurosurgery or spinal fusion.

Figure 2: Perioperative flowsheet for risk assessment(14).

Infective endocarditis prophylaxis:

Infective endocarditis is a rare condition but is difficult to treat and carries high morbidity and mortality rates, therefore preventative measures are essential. The European Society of Cardiology (ESC) and American Heart Association (AHA) advocates antibiotic prophylaxis in those with the highest risk of adverse outcomes.

High risk individuals in whom antibiotic prophylaxis should be provided are as follows(18):

- Patients with prosthetic valves or undergone valve repair using prosthetic material.
- Patients with a history of previous infective endocarditis.
- Patients with cyanotic congenital heart defects.
- Patients for the first six months after surgical or percutaneous repair of a congenital heart disease with a prosthetic material, and indefinitely in case of residual shunt or valvular regurgitation.

Procedure	Common Pathogens	Drug of choice	Dose(19)	Penicillin hyper-sensitivity	Dose(19)
Dental [a]	Viridans group streptococci	Amoxicillin Amoxicillin/ Ampicillin Second line Cephalexin Cefazolin/ Ceftriaxone	30mg/kg po 50mg/kg im/iv 50mg/kg po 50mg/kg im/iv	Clindamycin	20mg/kg
Implantable cardiac electronic device insertion	Staphylococci (Methicillin sensitive) Staphylococci (Methicillin resistant)	Cefazolin Vancomycin	50mg/kg iv 15mg/kg	Vancomycin	15mg/kg iv
Procedure in infected or colonised tissue					
Respiratory	Staphylococci	Cefazolin	50mg/k iv	Clindamycin Vancomycin	20mg/kg 15mg/kg iv
Genitourinary/ gastrointestinal	Enterococci	Amoxicillin/ ampicillin	30mg/kg im/iv/po 50mg/kg iv	Vancomycin	15mg/kg iv
Skin	Staphylococci Beta-haemolytic Streptococcus Staphylococci (Methicillin resistant)	Amoxicillin Cephalexin Cefazolin/ceftriaxone Vancomycin	30 mg/kg im/iv/ po 50mg/kg po 50mg/kg iv 15mg/kg iv	Clindamycin Vancomycin	20mg/kg iv 15mg/kg iv

Table 8: Summary of antibiotic use in endocarditis prophylaxis (18).

a. Antibiotic prophylaxis is recommended for invasive dental procedures that involve the manipulation of gingival tissue or periapical region or perforation of the mucosa when performed on high-risk individuals. According to the Australian guidelines the following dental procedures are the most likely to cause bacteraemia:

    - Tooth extraction.
    - Periodontal surgery, subgingival scaling and root planning.
    - Replantation of avulsed teeth.
    - Other surgical procedures such as implant placement or apicoectomy.

The rationale for restricting the use of prophylactic antibiotics lies in limiting the possibility of generation of antibiotic resistance with repeated use of these antibiotics, prevention of anaphylactic reactions, and limiting the cost burden associated with the cumulative cost of prescribing these antibiotics for a large number of patients(18).

Suggested anaesthetic goals for common CHD lesions:

Below is a suggested approach marrying the physiology of the cardiac lesion with the effects of pharmacological agents to maintain haemodynamic stability in the perioperative period. A crying and distressed patient will have sympathetic stimulation which increases oxygen consumption and myocardial work. This may not be well tolerated in a child with limited cardiac reserve(5). In this case an appropriate premedication should be considered.

Left to right shunt lesions:

VSD, ASD, PDA, AVSD:

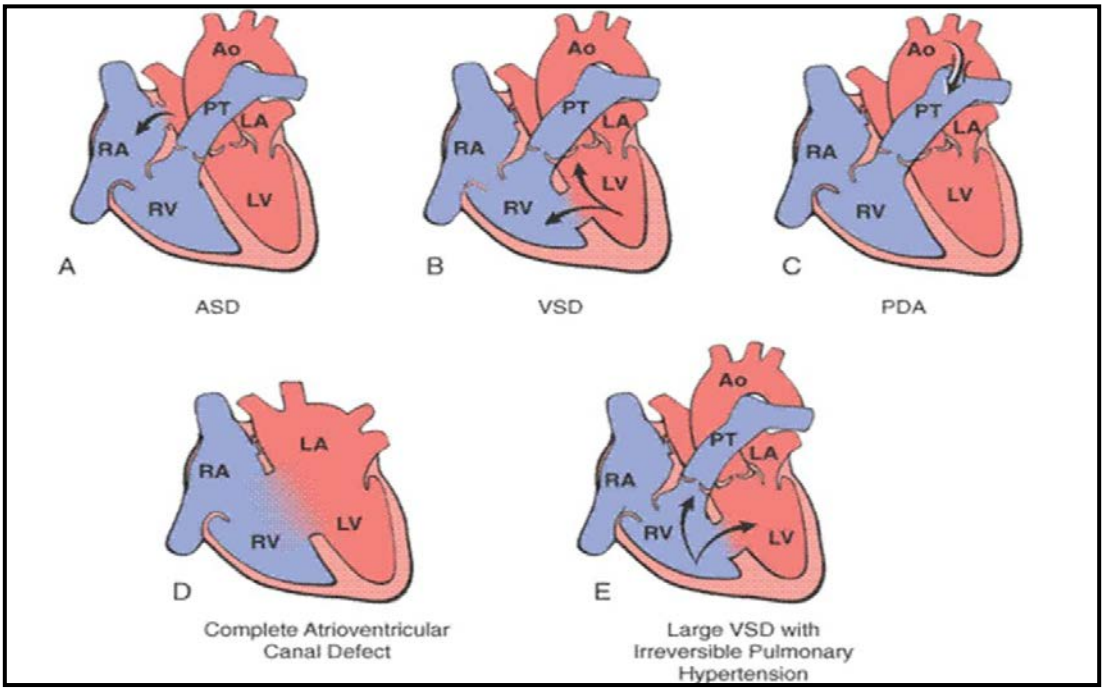


Figure 3: Left to right shunt lesions(20)

Manoeuvres that decrease PVR such O_2 administration, nitric oxide, low arterial CO_2, and alkalosis will increase the left to right shunt and pulmonary flow at the cost of decreased systemic flow(12). Shunt flow is influenced by the size of the lesion and the pressure gradient across the lesion.

ASD:

The defect and left to right shunt is at the atrial level and shunting occurs during diastole, while the atrioventricular valves are open(13). With increased pulmonary flow and venous return to the LA, there is bi-atrial volume overload and dilatation, and a progression to increased PVR. However, pulmonary hypertension is generally not seen in these patients until later. Oxygen saturation is higher in the RA compared to the SVC: step up in saturation. A higher saturation in the RA means a greater degree of shunt. Atrial pressures will be equal and normal, while RV pressure is lower than LV but slightly higher than normal, (about a third to half of systemic pressure in a large ASD) (12). A lesion less than 5mm is referred to as restrictive while the larger defects are non-restrictive and associated with atrial dilatation(13).

Goals:
Anaesthetic goals should include a slightly higher preload, slightly increased PVR to reduce pulmonary flow or a slight decrease in SVR. In an isolated lesion they usually do not pose a higher risk for infective endocarditis(13).

Methods
Inhalation induction or IM or IV ketamine are acceptable techniques(13).

VSD:

Here the defect and the left to right shunt are at the ventricular level, and shunting occurs during systole. The shunting during systole (higher pressures) means that it is haemodynamically more significant and progression to pulmonary vascular disease is sooner. Due to the increase in pulmonary circulation, venous return to LA and LV is higher and these are volume overloaded. The RV is not volume over-loaded as blood is preferentially directed to the low pressured pulmonary circulation via the pulmonary artery (PA). As a result there is a step up in saturation from the SVC to the PA. The pressure in the right ventricle is determined by the size of the defect. Larger defects (compared to the size of the aortic annulus) means higher RV pressures. In the face of pulmonary vascular disease RV pressure is increased(12). These patients develop symptoms earlier than in ASD and are more prone to infective endocarditis(13).

Goals:
Anaesthetic goals should again be to maintain a slightly higher preload and pulmonary vascular resistance, while keeping SVR slightly lower(13).

Methods
Inhalation induction or IM or IV ketamine are acceptable techniques. Higher PVR obligates the need to be ready to treat high PVR and right ventricular failure by use of inhaled NO, dobutamine and milrinone. Up to 10% of patients can develop conduction abnormalities after the VSD repair(13).

PDA:

The ductus arteriosus is the communication between the junction of the main and left pulmonary artery with the lesser curvature of the descending aorta after the origin of the left subclavian artery origin. PDA's are commonly seen in premature infants, females, at high altitudes or in association with maternal rubella(13).

Shunting is left to right during diastole and systole (continuous), because of the high aortic pressure. This creates a volume overload in the LA and LV, as a result of increased venous return and progression to pulmonary vascular disease. In a large PDA the diastolic shunting may produce steal of blood from coronary circulation. RV pressure is increased if there is pulmonary vascular disease(12).

Goals
The goals should be to limit the decrease in PVR by limiting FiO_2 and maintaining normocarbia.

AVSD:

AVSD's are also called common atrioventricular canal or endocardial cushion defect, resulting from the failure of endocardial cushions to fuse with the lower part of the atrial and upper part of the ventricular septums. The atrioventricular valves will also be abnormal. Due to the left to right

shunting and regurgitation from abnormal valves both ventricles and atria are overloaded, leading to pulmonary congestion and pulmonary hypertension(13).

Goals: Aim to maintain a lower degree of pulmonary resistance, using O_2, NO and analgesics. The anaesthetic technique should be tailored to the degree of pulmonary hypertension and left to right shunt(13).

		Pre-induction	Induction	Maintenance	Emergence
Systemic circulation	Rate	Avoid bradycardia			
	Rhythm	Maintain sinus rhythm			
	Contractility	Maintain or improve; consider positive inotropic agents. Avoid agents that cause negative inotropy.			
	Preload	Maintain by avoiding long starvation periods/ IV fluids	Slightly higher. Beware of fluid overload and worsening of congestive cardiac failure		
	Afterload	Slightly lower, may benefit from reduction.			
Pulmonary circulation	Slightly reduced flow is acceptable by either slight increase in PVR or decrease in SVR				
Ventilation	Ventilate early. Oxygenate well bearing in mind the effect of high O_2 concentrations on PVR.				

Table 9: Overview of anaesthetic goals in left to right shunt lesions(21).

Right to Left shunt lesions:

ToF:

The associated defects are labelled 1 to 4 in the diagram below. The right to left shunt is due to the RVOT obstruction(12), in the presence of a hypertrophied infundibulum (dynamic obstruction) or pulmonary stenosis (fixed obstruction)(13). The direction of the shunt means that pulmonary blood flow and venous return to the LV is decreased. The LV is therefore smaller than the RV. Oxygen saturation in the PA is equal to that in SVC, and the saturation in the aorta is less than in the pulmonary vein.

RV pressure is systemic and because of the stenosis, PA pressures are normal. In a ‘Tet” spell the RVOT obstruction worsens and all the blood from the RV is shunted across the VSD to the systemic circulation, therefore there is no pulmonary venous return to the left heart.

Goals: Avoid a low SVR, and desaturation at any time should be treated as a ‘Tet spell’ (13) provided that common causes such as a blocked/ kinked endotracheal tube are ruled out.

Methods: Gas induction can be done but with attention to maintain SVR with inotropes or vasopressors like phenylephrine. Ketamine induction also helps maintain SVR. Desaturation during a Tet spell should be treated with volume (increase RV preload), analgesics (decreases catecholamine release to decrease spasm), and increase SVR (vasopressors and compression on femoral arteries) to decrease right to left shunt (12, 13).

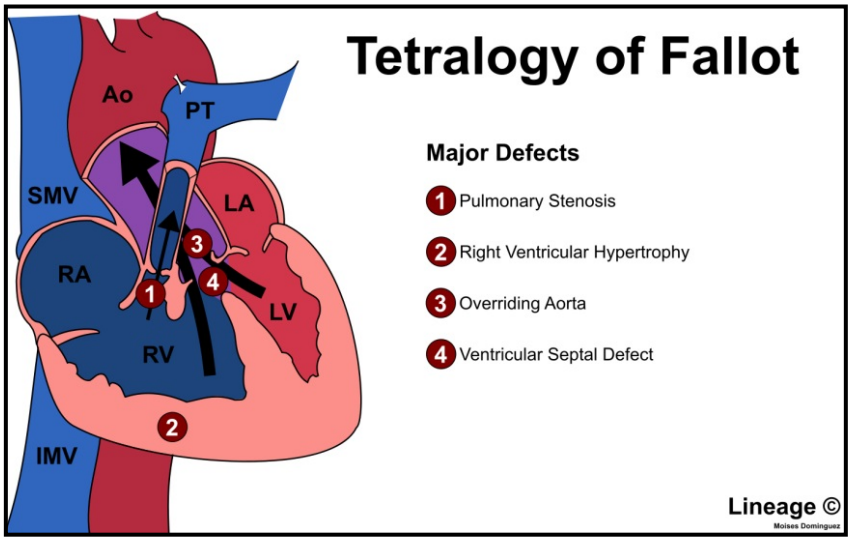


Figure 4: Tetralogy of Fallot (22).

TGA:

Here there is a parallel circulation and ventriculo-arterial discordance, where the RV is connected to the aorta and the LV is connected to the pulmonary artery. This means that physiologically there is a complete left to right shunt and complete right to left shunt. With deoxygenated blood from the RV is pumped back to the systemic circulation and oxygenated blood from the LV is pumped back to the lungs.

These patients will only survive if anatomically there exists another shunting lesion in the heart that allows some deoxygenated blood to reach the pulmonary system and oxygentated blood to reach the systemic circulation. These patients may have had an atrial sepostomy (palliation) done that which allows such a shunt to assist with treating the cyanosis and stabilisation before definitive surgery(12, 13). The condition is also associated with abnormal location of coronary arteries.

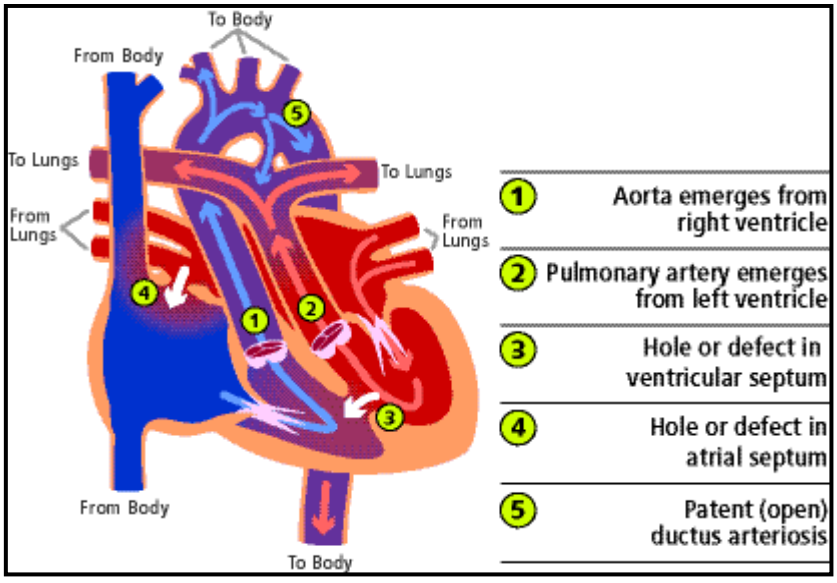


Figure 5: Transposition of great arteries(23).

Saturation data is difficult to interpret and is determined by the level and direction of shunt. RV pressures will be systemic and LV pressure will decrease in accordance with pulmonary pressures.
Goals:
Keep PVR lower than SVR and maintain cardiac output. Increased pulmonary flow in the face of lower PVR will allow more mixing of blood via shunts and improve saturation(13).

Methods:
Reduce pulmonary resistance by using:
Pharmacologic strategies: Nitric oxide, nebulised PGI_2, Sildenafil (oral/ IV)
Ventilatory strategies: Increased FiO_2, decreased $PaCO_2$, avoid high PEEP and use normal tidal volumes.
General strategies: avoid hypoxia, hypercarbia, acidosis, hypothermia, and hypoglycaemia maintain an alkalotic pH (13).

		Pre-induction	Induction	Maintenance	Emergence
Systemic circulation	Rate	Avoid a bradycardia			
	Rhythm	Maintain sinus rhythm, avoid arrhythmia triggers			
	Contractility	Maintain or improve and consider the use of inotropic agents			
	Preload	Maintain by avoidance of prolonged fasting periods (also assists in maintaining haematocrit)	Maintain or increased		
	Afterload	Slightly higher by avoiding hypovolemia, acidosis, hypoxia. Phenylephrine /ketamine assists by maintaining SVR.			
Pulmonary circulation	Decrease PVR in order to improve pulmonary circulation. Avoid triggers of infundibular spasm which will worsen right to left shunt: such as pain, anxiety crying, injury, sympathetic response or increased contractility. (24)				
Ventilation	Minimise intrathoracic pressures with normal to low ventilatory pressures, low PEEP and adequate neuromuscular blockade. This allows for adequate venous return and preload. Consider a decrease in inspiratory pressures and respiratory rate if there is a decrease in saturation. Avoid a high concentration of inspired O_2 as it may be harmful and will not improve saturation.				

Table 10: Overview of anaesthetic goals in right to left shunt lesions.

Common Palliative Procedures:

Certain CHD lesions may not be amenable to immediate, complete surgical correction. In these cases the aim will be to improve the baby's physiological state by e.g. controlling heart failure, improving cyanosis or preparing the circulation for later correction at a time more suitable to the babies age, weight or if immediate intra-cardiac correction carries a significantly higher mortality than staged shunting(25).

There are over 20 palliative procedures in total and these are classified into 4 classes depending on the goals and indications(25).

a. Increasing pulmonary artery flow for pulmonary oligeamia (includes shunt procedures)
b. Decreasing pulmonary artery flow for pulmonary over-circulation (Pulmonary artery banding and Norwood procedure)
c. Enhancing intra-cardiac blood-oxygen mixture for systemic hypoxemia (varying techniques for atrial sepostomy)
d. Other procedures: including congenital mitral or aortic stenosis palliation, co-arctation of aorta palliation and hybrid palliative procedures for HLHS.

This review will highlight a few of the commoner procedures.

Blalock –Taussig's shunt/ Modified Blalock-Taussig's shunt:

The above are performed as systemic to pulmonary artery shunting in order to establish pulmonary blood flow and relieve pulmonary venous obstruction. The indications therefore are in Tetralogy of Fallot, tricuspid atresia, pulmonary atresia with intact ventricular septum, pulmonary atresia with VSD, Ebstein's anomaly, single ventricle with pulmonary or aortic atresia and hypoplastic left heart syndrome.

The Blalock-Taussigs shunt is a direct connection between the subclavian artery and pulmonary artery. This allows blood flow down the pressure gradient to the pulmonary circulation. The disadvantages of this procedure such as long operative dissection time, phrenic nerve injury, technical difficulty during take down and possible arm ischemia, lead to the improved technique of the Modified Blalock- Taussig's Shunt. This procedure uses a graft (prosthetic or human vasculature) between the subclavian and pulmonary arteries(25).

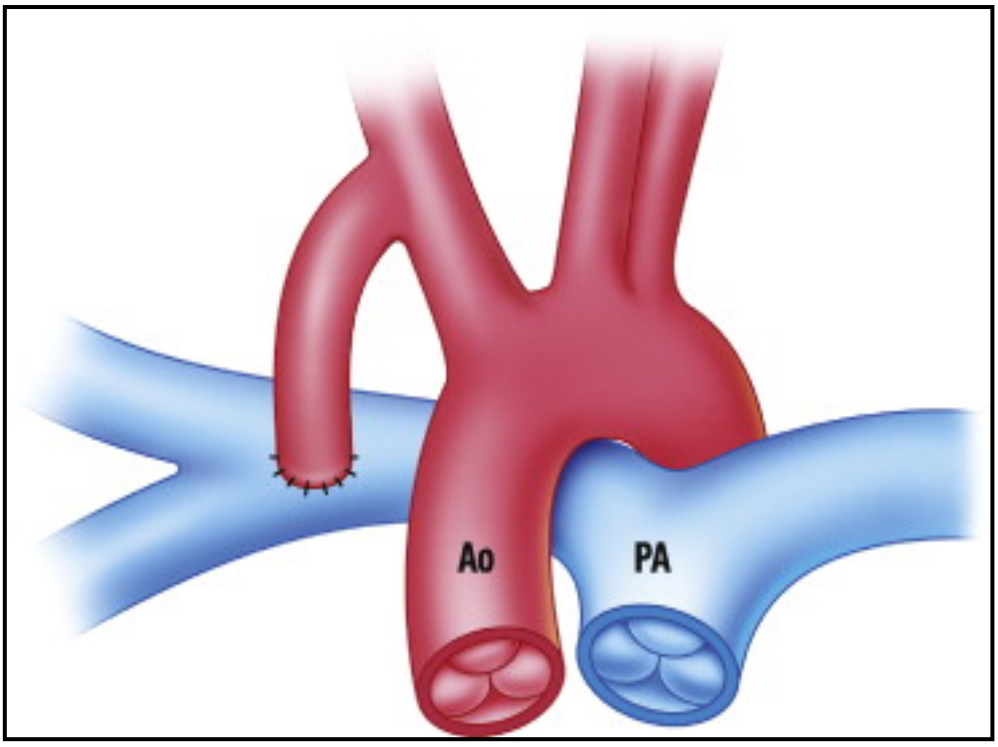


Figure 6: Classic Blalock-Taussig's shunt (Ao: Aorta, Pa:Pulmonary artery)(25).

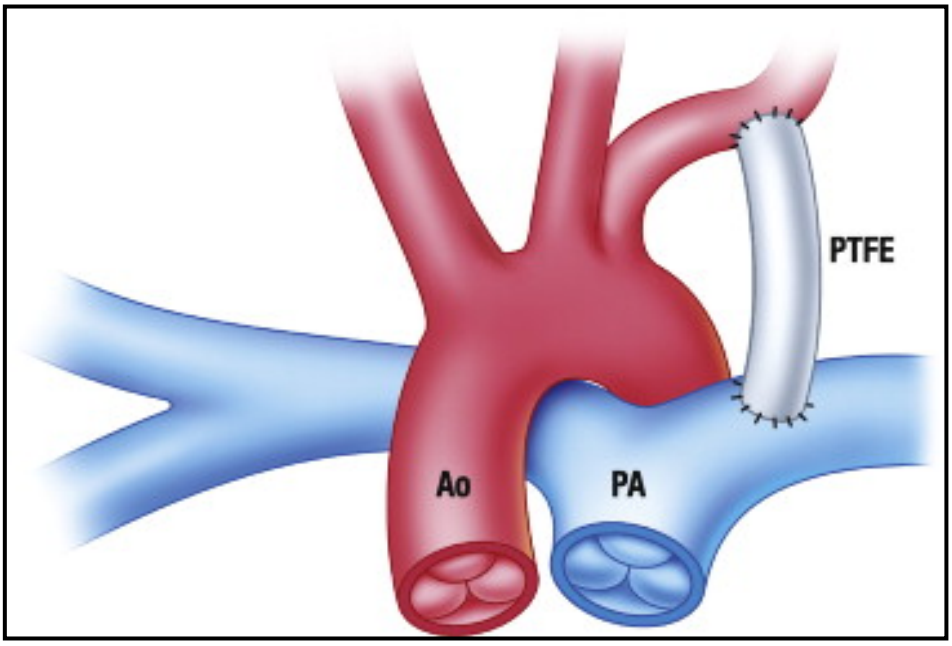


Figure 7: Modified Blalock-Taussigs Shunt
(Ao: Aorta, PA: Pulmonary Artery, PTFE: Polytetrafluorothene) (25)

'Wanna Be' Blalock-Taussig's shunt, named as such by Ruiz and Bailey, is actually a ductus arteriosus stent used in neonates and infants with duct dependant cyanotic congenital heart disease. It allows the team to gain time for the child and pulmonary arteries to grow while leaving the surgical field for definitive surgery untouched(25).

Glenn's shunt

Used in a diverse range of cyanotic congenital heart disease. It relieves the ventricle of volume and hence work overload while providing or improving venous blood flow to the lungs instead of an arteriovenous mixture. Thus avoiding the possibility of failure of a Fontan circulation (discussed under the next heading) while partially relieving symptoms(25). It is often used as palliation or an intermediate procedure for hearts with only one usable ventricle e.g. tricuspid atresia(12).

The classic Glenn Shunt is a connection between the SVC and the distal right pulmonary artery while the proximal end is tied off. The bidirectional Glenn shunt is done by connecting the SVC to the right branch of the pulmonary artery and allowing flow to both lungs. This is also called hemi-Fontan as it is equivalent physiologically to half a Fontan shunt. The Glenn shunt is a step towards a Fontan circulation in a single ventricle circulation(25).

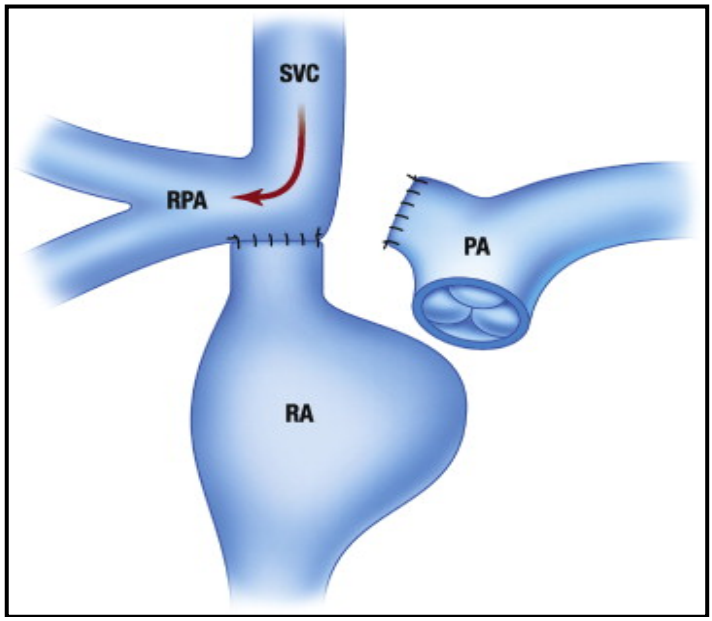


Figure 8: Classic Glenn's Shunt (SVC: superior vena cava, RPA: right pulmonary artery, PA: pulmonary artery, RA: right atrium) (25).

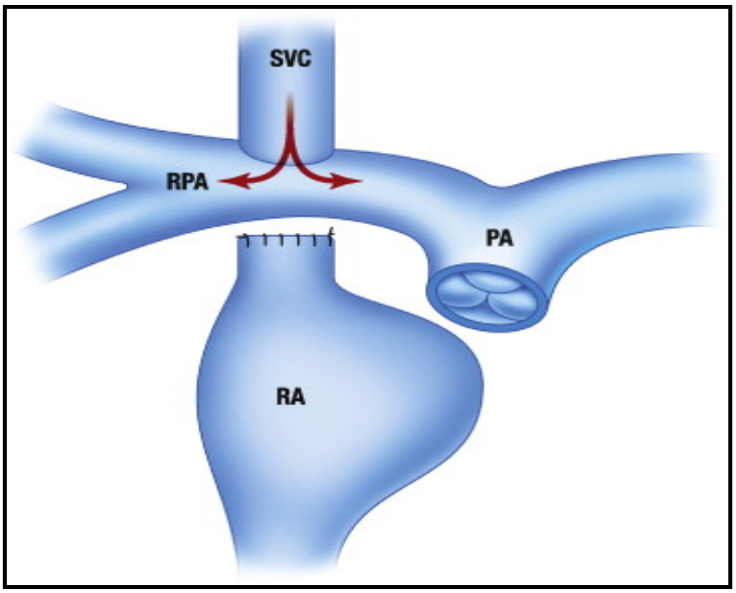


Figure 9: Bidirectional Glenn's Shunt(SVC: superior vena cava, RPA: right pulmonary artery, PA: pulmonary artery, RA: right atrium) (25).

Importantly, there is no pumping chamber incorporated and blood flow to the lungs is passively down a pressure gradient. The circulation depends on upper body circulation (especially brain) and distally on the PVR. Any manoeuvre that decreases cerebral venous return will decrease flow in the Glenn shunt and hence venous perfusion to the lungs e.g. Hyperventilation. Therefore a balance in ventilation is required to avoid extreme situations, keeping in mind that hyperventilation also decreases PVR and augments pulmonary flow(12).

Fontan circulation:

Here both the SVC and the IVC are directed passively to the pulmonary circulation, without a pumping chamber. Therefore pulmonary circulation again relies on low PVR. An extra-cardiac (outside the heart) Fontan Baffle minimizes atrial surgery and future arrhythmia risk. A fenestration can be created between the Fontan Baffle and the right atrium to allow a 'pop-off' when pressure within the baffle is high. This maintains cardiac output but at the cost of cyanosis.

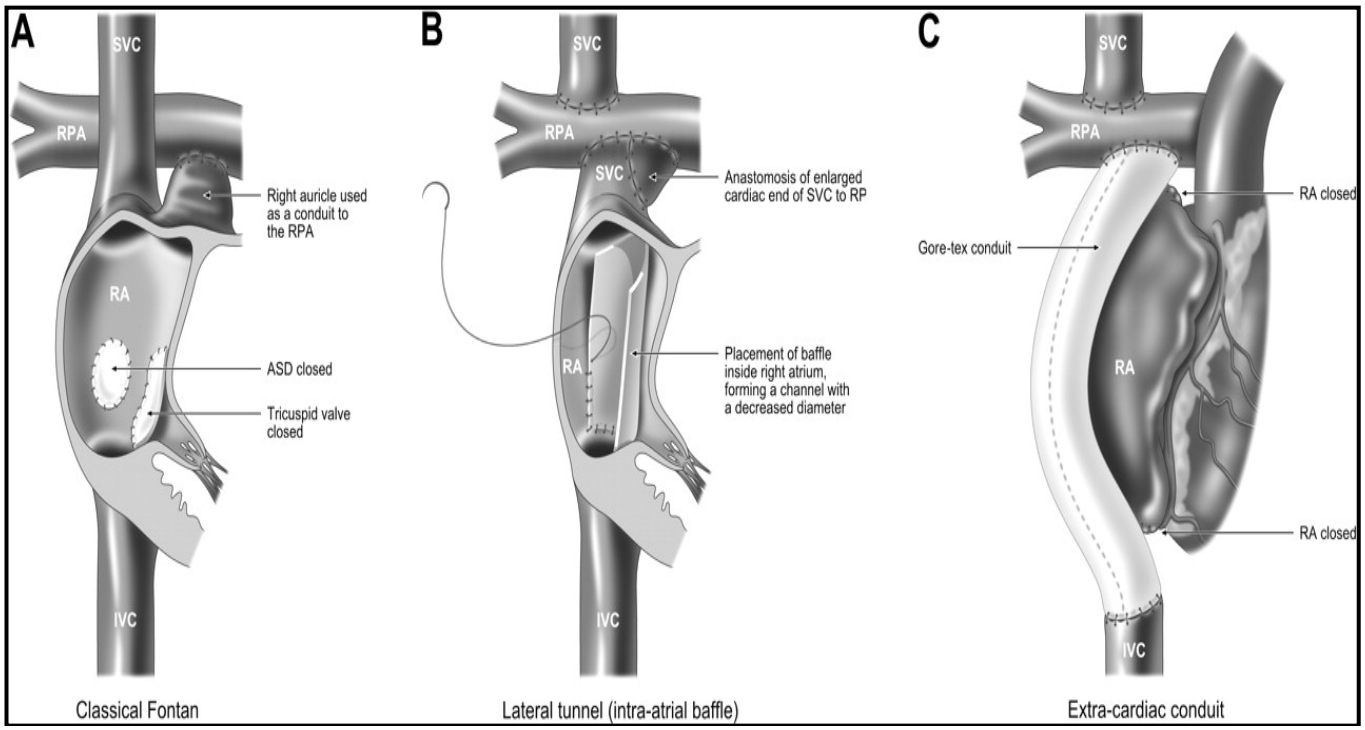


Figure 10: Fontan surgical techniques: Classical atrio-pulmonary connection (A), Lateral tunnel (B), and extra-cardiac conduit (C) (IVC: inferior vena cava, SVC: superior vena cava, RPA: right pulmonary artery, PA: pulmonary artery, RA: right atrium) (26).

Positive pressure ventilation increases intra-thoracic pressures and hinders the Fontan circulation necessitating the termination of positive pressure ventilation as soon as possible. Hyperventilation will have the same effect(12).

Norwood Procedure:

This procedure is typically used in palliation of HLHS but can be used in other conditions. Some of these are: aortic atresia or stenosis with inadequate left ventricle, mitral atresia or stenosis, interrupted aortic arch, double outlet right ventricle with mitral atresia, or complete transposition with hypoplastic right ventricle(25).

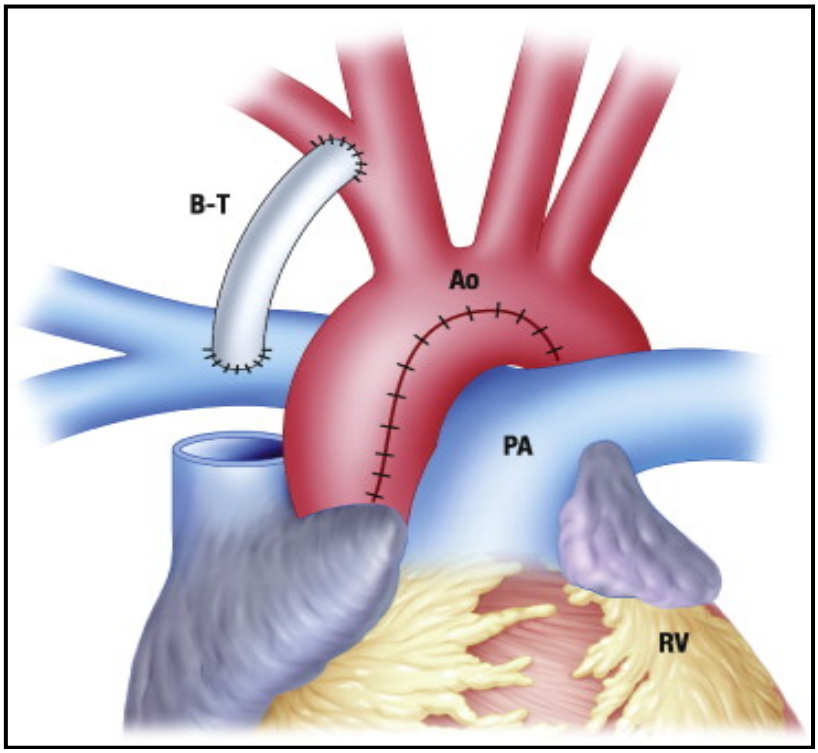


Figure 11. Norwood Procedure with a Blalock Taussig's Shunt
(B-T: Blalock Taussig's shunt, Ao: aorta, PA : pulmonary artery, RV: right ventricle) (25)

The procedure includes a neo-aorta reconstruction and aortic arch augmentation(25). It is the first stage of palliation in HLHS and patients are still dependant on the fragile single ventricle for both systemic and pulmonary circulation(14). The second and third stages of treatment would then include a Glenn's shunt and fontan circulation respectively. These restore the series circulation and reduces the volume overload on the single ventricle(14).

Conclusion

It is evident that CHD disease encompasses a vast array of conditions/ lesions associated with complex cardiac and respiratory physiology. Important pre-operative, intraoperative and post-operative consequences of their CHD have significant impact on their morbidity and mortality(14).

For safe anaesthetic management of these patients it is essential to understand the pathophysiology of these lesions, the physiology of corrective procedures while keeping in mind the physiological state of the patient presenting for a non-cardiac procedure. Early preparation and multidisciplinary care is integral in managing these children.

Annexure 1

ACS	American College of Surgeons
ALCAPA	Anomalous left coronary artery from pulmonary artery
AS	Aortic stenosis
ASD	Atrial septal Defect
AVSD	Atrioventricular septal defect
CHD	Congenital heart disease
CoA	Co-arctation of the Aorta
HLHS	Hypoplastic left heart syndrome
HRHS	Hypoplastic right heart syndrome
IVC	Inferior vena cava
LA	Left atrium
LV	Left ventricle
NSQIP	National Surgical Quality Improvement Programme
PAP	Pulmonary artery pressure
PaCO2	Partial pressure of carbon dioxide
PaO2	Partial pressure of oxygen
PEEP	Positive end expiratory pressure
PDA	Patent ductus arteriosus
PS	Pulmonary stenosis
Q	Flow
Qp	Pulmonary flow
Qs	Systemic flow
RA	Right atrium
RV	Right ventricle
RVOT	Right ventricular outflow tract
SVC	Superior vena cava
TA	Truncus arteriosus
TAPVC	Total anomalous pulmonary venous connection
TGA	Transposition of Great Arteries
ToF	Tetralogy of Fallot
VSD	Ventricular septal defect

Annexure 2

References

1. White MC. Approach to managing children with heart disease for noncardiac surgery. Pediatric Anesthesia. 2011;21(5):522-9.
2. van der Linde D, Konings EE, Slager MA, Witsenburg M, Helbing WA, Takkenberg JJ, et al. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. Journal of the American College of Cardiology. 2011;58(21):2241-7.
3. Storey E. RECOGNISING CARDIAC DISEASE IN CHILDREN ANAESTHESIA TUTORIAL OF THE WEEK 2008, 26 May.
4. Hoosen E, Cilliers A, Hugo-Hamman C, Brown S, Lawrenson J, Zuhlke L, et al. Paediatric cardiac services in South Africa. SAMJ: South African Medical Journal. 2011;101(2):106-.
5. Menghraj SJ. Anaesthetic considerations in children with congenital heart disease undergoing non-cardiac surgery. Indian J Anaesth. 2012;56(5):491.
6. White MC, Peyton JM. Anaesthetic management of children with congenital heart disease for non-cardiac surgery. BJA Education. 2011;12(1):17-22.
7. Faraoni D, Zurakowski D, Vo D, Goobie SM, Yuki K, Brown ML, et al. Post-operative outcomes in children with and without congenital heart disease undergoing noncardiac surgery. Journal of the American College of Cardiology. 2016;67(7):793-801.
8. Ramamoorthy C, Haberkern CM, Bhananker SM, Domino KB, Posner KL, Campos JS, et al. Anesthesia-related cardiac arrest in children with heart disease: data from the Pediatric Perioperative Cardiac Arrest (POCA) registry. Anesthesia & Analgesia. 2010;110(5):1376-82.
9. Lee C. Transitional Physiology. FCA Part 1 Anaesthetic refresher course2016.
10. Butterworth JF, Mackey DC, Wasnick JD. Morgan and Mikhail's clinical anesthesiology: McGraw-Hill Education; 2018.
11. Foetal circulation [Available from: (https://i.pinimg.com/originals/cf/d0/ac/cfd0ac181daf3af4326d3ae75bf4f32f.jpg Diagram)
12. Chowdhury D. Pathophysiology of congenital heart diseases. Annals of cardiac anaesthesia. 2007;10(1):19.
13. Hamid M. Anaesthetic considerations for congenital heart disease patient. Perioperative Considerations in Cardiac Surgery. 2012:57-72.
14. Pilkington M, Egan JC, editors. Noncardiac surgery in the congenital heart patient. Seminars in pediatric surgery; 2019: Elsevier.
15. Friesen RH, Williams GD. Anesthetic management of children with pulmonary arterial hypertension. Pediatric Anesthesia. 2008;18(3):208-16.
16. Ramakrishna G, Sprung J, Ravi BS, Chandrasekaran K, McGoon MD. Impact of pulmonary hypertension on the outcomes of noncardiac surgery: predictors of perioperative morbidity and mortality. Journal of the American College of Cardiology. 2005;45(10):1691-9.
17. Hennein HA, Mendeloff EN, Cilley RE, Bove EL, Coran AG. Predictors of postoperative outcome after general surgical procedures in patients with congenital heart disease. Journal of pediatric surgery. 1994;29(7):866-70.
18. Kaya CT, Erol, C. How to Achieve Infective Endocarditis Prophylaxis Ankara University School of Medicine, Cardiology Department, Ankara, Turkey2018 [12 December 2018:[Available from: https://www.escardio.org/Journals/E-Journal-of-Cardiology-Practice/Volume-16/vol16no33.
19. Taylor JL. Paediatric Anaesthesia Rotation Orientation Booklet. 2019.
20. Cram.com. [updated 2013, September. Available from: https://www.cram.com/flashcards/anatomy-of-congenital-heart-disease-3404398.
21. Thomas JM. Anaesthesia for congential heart disease: Pointers and pitfalls Continuing Medical Education, [Sl]. 2011;29(11/12):463.
22. Medbullets.com. [updated 2019, January 27. Available from: https://step2.medbullets.com/cardiovascular/120002/tetralogy-of-fallot.
23. mottchildren.org. 2019 [Available from: https://www.mottchildren.org/conditions-treatments/ped-heart/conditions/transposition-great-arteries.

24. Bester K. An approach to the cyanosed child for anaesthesia. Part II Anaesthesia Refresher Course 20122012.
25. Yuan S-M, Jing H. Palliative procedures for congenital heart defects. Archives of cardiovascular diseases. 2009;102(6-7):549-57.
26. d'Udekem Y, Iyengar AJ, Cochrane AD, Grigg LE, Ramsay JM, Wheaton GR, et al. The Fontan procedure: contemporary techniques have improved long-term outcomes. Circulation. 2007;116(11_supplement):I-157-I-64.