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Review Article

Grown up Congenital Heart Disease patient presenting for non cardiac surgery: Anaesthetic implications

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Abstract

Congenital heart disease patients surviving to adulthood have increased over the years due to various reasons. These patients are admitted in the hospital for non cardiac surgeries and other procedures more often than normal adult population. Management of grown up congenital heart disease patient presents a challenge during perioperative period for cardiologists, surgeons, intensivists and particularly for the anaesthetist. Management issues include psychological and physiological impact on the patient, complexity of defects, presence of previous palliative procedure, impact of anaesthetic agents on shunting and myocardium, endocarditis prophylaxis and associated extra cardiac anamolies.

Introduction

Population of adults with congenital heart disease (CHD) has increased over the years[1] due to improvement in paediatric cardiology, improved surgical and anaesthetic techniques[2] and better postoperative care. Expectations are that soon there will be more adults than children with CHD who have undergone some sort of palliative or corrective surgery.[3] Incidence of CHD is about 0.8% of all live births[4] and 85% of these CHD patients are expected to survive to adulthood in USA.[5] Early surgical intervention has also improved their chances of survival[6] and reduces complications associated with heart defects.

Presence of grown up congenital heart (GUCH) disease poses increased risk of mortality and morbidity under anaesthesia.[7] Anaesthetic management of these patients particularly the uncorrected group, in the operating room is challenging in several respects. Firstly, some heart defects are so complex that involvement of paediatric cardiologists and intensivists is necessary for complete understanding of the anatomy and pathophysiology. Additionally the management is quite complex and the anaesthetist needs to make an individualized anaesthetic plan[8] after several considerations. These essentially look at the effect of anaesthetic drugs on the heart and shunt, fluid management, effect of ventilatory changes on shunts and how to avoid pulmonary hypertension.

Most of these CHD defects can be categorized into those associated with increased pulmonary flow, reduced pulmonary flow and obstructive lesions. Other factors which should be considered are age of the patient, cardiac lesion, previous surgery performed (palliative or corrective), presence of cardiac complication and associated congenital anomalies.

Preoperative Considerations:

GUCH patients coming to preoperative clinic can be

grouped into three categories: Non operated patient; patient with previous palliative surgery[9] and patient with previous corrective surgery. Patients with total corrective surgery may still have residual defects. These patients may have single ventricle physiology, single RV or complex intra cardiac baffles.

Detailed information should be obtained about cardiac lesion, altered physiology and its implications under anaesthesia. Information about the age is very important as some lesions require early repair. Delayed surgery may otherwise lead to complications like pulmonary HTN, poor development of pulmonary vessels and failure to gain weight, which indicate cardio respiratory decompensation. Poor exercise tolerance is indicated by fatigue and dyspnoea on feeding, irritability and failure to gain weight. Previous cardiac and non cardiac surgeries and prolonged intubation should be enquired as they suggest subglottic stenosis.

Cyanosis and congestive heart failure (CHF) are major manifestations of CHD. Cyanosis occurs due to decreased pulmonary flow anatomically or functionally (Mixing lesion). Cyanosis may be permanent or appears intermittently. Central cyanosis is recognizable when deoxygenated haemoglobin in arterial blood is > 3 gm/dl.

Along with left ventricular function, the right ventricular function should also be assessed as it is equally important in the paediatric CHD patient. Patients with high pulmonary flow may present with tachycardia, tachypnoea, irritability, cardiomegaly and hepatomegaly. History of wheezing, frequent respiratory infection and pneumonia is also common. The amount of cardiac reserve is assessed by exercise tolerance in older children. Pregnant patients with GUCH require special attention on preoperative evaluation. These patients need regular follow up, early involvement of anaesthetist, assessment of degree of cardiovascular impairment and optimization of pulmonary vascular resistance. High foetal mortality[10] is seen in mothers particularly with low saturation and very high haematocrit levels.

Associated non cardiac congenital anomalies include musculoskeletal abnormalities 8.8%, neurologic defects 6.9% and genito urinary irregularities 5.3%. Downs syndrome patients may have atlanto -occipital subluxation.

Drug history can show the use of warfarin, antidepressants, diuretics and anti arrhythmics with their associated side effects. Laboratory investigation should be tailored accordingly.

NPO orders should be clearly written with timing if possible. Dehydration should be avoided in cyanotic GUCH patients. If timing of surgery is uncertain then an IV line should be placed and fluids started.

Midazolam[11] is the preferred sedative to reduce oxygen consumption in the doses of 0.5 mg/kg orally half hour before surgery. If IV is present then incremental doses of 0.1 - 0.25 mg midazolam can be given.

Endocarditis prophylaxis has recently been revised[12,13] For dental procedures AHA recommend prophylaxis in patients is a follows:

◆ When gingival tissues manipulated, or periapical region of teeth or perforation of oral mucosa

◆ Prior history of infective endocarditis

◆ Non-repaired cyanotic congenital heart disease (CHD), including shunts and conduits

◆ Complete CHD repair within the previous six months

◆ Repaired CHD with residual defects

Antibiotic for infective endocarditis prophylaxis is no longer indicated in patients with

◆ Aortic stenosis, mitral stenosis, or symptomatic or asymptomatic mitral valve prolapse.

◆ Genitourinary and gastrointestinal tract procedures (transesophageal echocardiography, esophagogastroduodenoscopy, colonoscopy, etc.) do not warrant infective endocarditis prophylaxis unless active infection is present.

Investigation:

Complete blood count and coagulation profile should always be checked. Polycythaemia[14] increases blood viscosity which leads to thrombosis and infarction in cerebral, renal and pulmonary region. PT and PTT are usually abnormal in the polycythaemic patient. Coagulation abnormalities also occur due to platelet dysfunction, hypofibrinogenaemia and factor deficiencies. Preoperative phlebotomy[15] is performed in symptomatic hyperviscosity and HCT > 65%. Dehydration may further aggravate symptoms and should be corrected before deciding about phlebotomy. WBC count and CRP gives clue to the diagnosis of infection.

Serum electrolytes should be checked in patients receiving diuretics. Hypocalcaemia is commonly found in patients with Di George syndrome. Recent ECG, ECHO and catheterization findings are very important to decide about anaesthesia management. ECG may show ventricular strain or hypertrophy. ECHO used for doppler and colour flow mapping while catheterization is used for information about pressures in different chambers, magnitude of shunt and coronary anatomy.

The chest X-Ray shows the heart position (Dextrocardia) and size, atelectasis, acute respiratory infection, vascular markings and elevated hemidiaphragm. Patients with diminished pulmonary blood flow show reduced pulmonary markings.

Intraoperative Considerations:

All intravenous tubings, free of air bubble and preferably filters,[16] should be placed in patients with Eisenmenger's syndrome. Hypothermia should be avoided and. Polycythaemic patient must be well hydrated[17] before induction either by IV or orally. All commonly used induction agents are well tolerated depending on the rate and dose of the drug. Reduction in SVR should be considered when using intravenous agents. Inhalation induction is acceptable in GUCH patients with simple cardiac lesion. Patients with poor cardiac function, who requires ionotropes preoperatively, may not tolerate inhalation induction. Ionotropes should be continued and IV induction agents titrated. Decision about invasive monitoring should be individualized according to the type of surgery and cardiac lesion. Internal juglar vein cannulation may pose a risk of thromboembolism in patients with Fontan circulation. In addition pulmonary artery catheter placement may be difficult in patients with anatomical abnormalities and may be misleading in patients with intra cardiac shunt. Use of ECHO and blood gases may provide useful information instead of pulmonary artery catheter.

Intraoperative Management:

Depends on presence of shunt, pulmonary HTN, hypoxaemia. Ventricular dysfunction, pulmonary flow and arrhythmia.

Shunt:

These are abnormal communications (congenital or surgically created) between heart chambers or blood vessels. Blood flow through shunt depends upon diameter of defect and balance between systemic and vascular resistance. Balance between SVR and PVR is essential in the anaesthetic management of patient with shunts.

Left to Right Shunt (ASD, VSD, PDA, AV canal defects, PAVD, BT shunt):

It has minimal effect on inhalation or intravenous induction and decreases with the drop in SVR or an increase in PVR. Left to right shunts leads to excess pulmonary blood flow. Patients are acyanotic but deterioration in gas exchange may result from pulmonary congestion. 1-1.5 MAC of Isoflurane, halothane and sevoflurane has no effect on Qp: Qs ratio in patients with isolated ASD or VSD during mechanical ventilation.[18] Remember High PaO_2 and low $PaCO_2$ have pulmonary vasodilating properties which in turn may further increase pulmonary congestion. 100% oxygen and hyperventilation in patients with L-R shunt should be avoided.

Patients with PDA are more vulnerable to coronary ischaemia[19] due to ongoing pulmonary runoff during the diastolic phase of the cardiac cycle and potential for low diastolic blood pressure. Therefore DBP should be monitored during surgery. Modified Blalock Taussig shunt (BT shunt) is an artificial shunt between subclavian artery (usually right) and pulmonary artery. Its dimension 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 pressure monitoring in the ipsilateral arm should be avoided.

Right to Left:

Intra cardiac shunts prolong inhalation induction while IV induction is faster. R-L shunt (e.g. TOF) or shunt reversal occur when SVR drops or PVR increases.[20] Increase R-L shunting leads to drop in SaO_2 which will not respond to increasing inspired oxygen concentration. Dead space ventilation will be increased due to reduced pulmonary flow. Hypercyanotic spell under anaesthesia responds to volume, Increase SVR with alpha agonists such as Phenylephrine and decreasing right ventricular outflow track obstruction with beta blockade.

Hypoxaemia:

Uncorrected or partially palliated GUCH patients may present with hypoxaemia. Two mechanisms are basically responsible for hypoxaemia: Inadequate pulmonary blood flow and/or admixture of deoxygenated with oxygenated blood in systemic circulation. These patients are polycythaemic with its associated complications. Dehydration should be avoided at all costs.

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

Those patients in whom pulmonary flow is normal and hypoxaemia is due to mixing of pulmonary and systemic blood, the management is more complicated. Pulmonary vasodilation in these patients increases cardiac work and decreases systemic blood pressure. Normal SaO_2 in these patients should not be expected and myocardial contractility and balance between systemic and pulmonary flow should be maintained.

Pulmonary Hypertension:

Mean pulmonary artery pressure greater than 25 mm Hg at rest is defined as Pulmonary hypertension (PHTN). High pulmonary flow as occurs in unrestricted L-R shunt will lead to CHF and pulmonary HTN. Initially 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 (PVR) exceeds SVR and symptoms appear due to R ⇌ L shunt, is the Eisenmenger syndrome.[13]

Eisenmenger Syndrome:

This is the commonest reason for cyanosis in GUCH patients. These patients have very high PVR (> 800 dynes cm^{-5})[21] and reversal or bidirectional intracardiac shunt. Most of these adults have simple congenital cardiac lesion which has progressed to severe pulmonary HTN while patients with complex lesions usually manifest with this syndrome early in life. Eisemenger patients usually have hypoxaemia, myocardial dysfunction and arrhythmias.

Anaesthetic risk is quite high even for minor surgeries[22] including cardiac arrest and pulmonary hypertensive crisis.[23] In addition, these patients are high risk for bleeding due to platelet dysfunction, thrombosis due to Polycythaemia, paradoxical embolism and arrhythmia. Preoperative phlebotomy is indicated in patients with hyperviscosity syndrome. Intravenous fluid should be administered in preoperative fasting phase to avoid hypotension episodes. Anaesthetic management focused on preventing further increase in R-L shunt by keeping SVR high and PVR low, maintaining myocardial contractility and prevention of arrhythmia and hypovolumia. General anaesthesia is preferred by most anaesthetists due to fear of reduced SVR by regional anaesthesia. Regional anaesthesia can be used[24,25] but SVR should be maintained at all cost. Watch for ventricular and supraventricular tachycardia in the postoperative phase and keep them in ICU or high dependency unit.[26] Pregnancy carries a very high risk of mortality and should be avoided.

Ventricular Dysfunction and Arrhythmias:

Impaired ventricular function[20] occurs due to volume overload (Large shunts, valvular regurgitation), Obstructive conditions and cardiac muscle diseases. Patient may present with tachycardia, tachypnoea, pulmonary congestion, hepatomegaly, gallop rhythm, hypotension, and decrease capillary refill. Arterial blood gas and X-Ray may show metabolic acidosis and pulmonary oedema respectively.

Elective surgery should be postponed in patients with signs and symptoms of cardiac failure for optimization. Patient may require digoxin, diuretics and ionotropes. Mechanical ventilation may be necessary for pulmonary oedema. Etomidate and fentanyl provide cardiovascular stability at the time of induction. Intravenous drugs take longer time to reach target areas due to prolonged circulatory time in patients with cardiac failure. Limit the use of inhalation anaesthetics due to associated myocardial depression. Maintain normal sinus rhythm and preload during anaesthesia. After load reduction by vasodilators may be required in many situations to reduce cardiac workload and improve cardiac output.

Arrhythmias are very common in GUCH due to scarring induced by previous cardiac surgery. In addition to that over distension of atria or ventricular can also lead to arrhythmia.[27] Supraventricular arrhythmias are more common than ventricular and may not respond to medical management. Anaesthetist has to deal with these patients when they come for radiofrequency ablation or surgical intervention. TOF related ventricular tachycardia is more manageable with antiarrhythmics.

Pregnancy:

Physiological changes during pregnancy pose great danger to the life of CHD patients.[28] These changes include 40 - 50% increase in blood volume, decrease in systemic and pulmonary vascular resistance and an increase in cardiac output. In addition, all cardiac chambers dimension increases particularly the right sided chambers.[29] This enormous volume loading on the heart can easily lead to cardiac failure (Incidence 4.8%)[10] and arrhythmias. Most common arrhythmias are supraventricular tachyarrhythmias (4.5%)[10] particularly in complex CHD. Four predictors of cardiac event during pregnancy[30] are mentioned (1) Prior cardiac event or arrhythmia (2) Baseline NYHA class >II or cyanosis (3) Left sided heart obstruction (4) Decreased systemic ventricular function.

Multidisciplinary team of an obstetrician, cardiologist, anaesthesiologist and neonatologist is mandatory to take care of these patients. Counseling should be started at an early age by paediatrician as unplanned pregnancy can be life threatening for patients with CHD. Once pregnancy is established then regular follow ups and team approach can save the patient's life. Termination of pregnancy should be considered when patient is in cardiac failure. Obstetrical complications include hypertension, thromboembolism and heart failure. Previously C-Sections under general anaesthesia was considered to be the best option. But now the use of epidural catheter with low dose of slow titrated local anaesthetics is more common for labour and delivery. Risk of reduction in SVR can be avoided by the use of intrathecal or epidural narcotics.

Invasive monitoring should be done according to the type of defect and functional status of patient. Arterial line should be placed below the obstruction in patients with coarctation to assess placental flow. An inline air filter can be used in patients with right to left shunts. Trendlenberg and lithotomy position[31] in Glenn shunt, Fontan and other cavopulmonary shunts leads to increased central venous pressure and compromises cerebral perfusion.

Oxytocin should be given at lowest effective dose and in infusion form. Increased uterine contraction due to oxytocin can easily lead to heart failure as each contraction pushes 300-500 ml blood into circulation. Warfarin crosses the placenta[16]

and has teratogenic effect while heparin is quite safe and most of these patients will be on low molecular weight heparin (LMWH). Most thromboembolic deaths occur during postpartum period for which LMWH need to be continued.

Postoperative Care:

High risk patients should be observed either in the intensive care unit or a monitored bed.[17] Arrhythmias and cardiac ischaemia in the postoperative phase should be observed. Adequate analgesia should be provided to the patients to prevent haemodynamic instability. Fluid balance is very important in these patients. Too much fluid can lead to cardiac failure in poor ventricular function while too little fluid causes hypotension.

Spontaneous ventilation is preferred and should be resumed as soon as possible in the postoperative period. Every effort should be made to reduce pulmonary pressure in ventilated patients by avoiding atelectasis, acidosis and by reducing the duration of inspiration.

Abbreviations: Key

CHD: Congenital Heart Disease. GUCH: Grown up Congenital Heart Disease. Qp-Qs: Pulmonary to Systemic Blood Flow Ratio. PaO2: Partial Pressure of Oxygen in Blood. PaCo2: Partial Pressure of Carbondioxide in Blood. MAC: Minimal Alveolar Concentration. ToF: Tetralogy of Fallot. ASD: Atrial Septal Defect. VSD: Ventricular Septal Defect. PDA: Patent Ductus Arteriosus.

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