

The Grown up Congenital Heart Disease Patient for Incidental Surgery.

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Introduction

In the United States alone it is currently estimated that there are between 320,000 and 600,000 adults with moderate or complex congenital heart disease (CHD).(1,2) This patient population group is growing rapidly and requires ongoing cardiological care.(1,2) The implication of CHD for these patients is profound. For example, life insurance policies and mortgages are refused in 37% of patients with CHD vs 6% of patients without.(3) Employment prospects are not always good, genetic counseling may be required, pregnancy may not be possible and exercise tolerance may be severely limited.(4,5) These patients present for multiple medical procedures requiring surgery and anaesthesia. Currently, there is a shortage of specialized training centers and cardiologists to take care of the growing population of adults with CHD.(1) Very often the anesthesiology provider will be called upon to determine if these patients are medically optimized and able to safely undergo incidental surgery. The answer to this question, especially in patients with cyanotic or complex CHD, will require an understanding of the pathophysiology of CHD, the physiology of single ventricle anatomy and consultation with respiratory physicians and adult cardiologists with a special interest in pediatric CHD.

This lecture will focus on the pathophysiology of CHD and how this influences anesthetic principles.

Although it may be simpler to think of CHD as uncorrected or surgically corrected, there are six pathophysiological subgroups (Table 1) of patients with CHD which may present for anaesthesia and incidental surgery. The more complex the diagnosis and surgery, the more thorough the examination and special investigations need to be.

Table 1.

Pathophysiological	Example
Uncorrected Simple CHD	<i>ASD, Restrictive VSD</i>
Surgically Corrected Simple CHD 2 Ventricle Repair	<i>ASD, VSD, PDA, Tetralogy of Fallot, Coarctation of the aorta</i>
Surgically Corrected Complex CHD 2 Ventricle Repair	<i>Transposition of the great vessels, Anomalous Pulmonary Venous Drainage, Truncus Arteriosus</i>
Surgical palliation for Single ventricle anatomy	<i>HLHS, HRHS, Tricuspid Atresia, Pulmonary Atresia, Heterotaxy Syndromes</i>
Uncorrected Complex CHD	<i>Pulmonary hypertension, Eisenmenger's Syndrome</i>
Heart Transplant for severe CHD	

Surgically corrected CHD:

Firstly establish the type of cardiac surgery that has been completed. Has a full, normal anatomical correction, without residual defects been achieved for the cardiac lesion? If so, these cases may be without significant added complexity for the anesthesiology provider.

Importantly however, without ongoing regular cardiological surveillance the following complications of surgically corrected congenital heart disease may commonly develop in adolescent and adult patients (Table 2.); It should be realized that many of the more severe complications are more likely to be present with surgically corrected complex congenital heart disease that requires multiple cardiac surgeries and patients with single ventricle physiology that despite numerous surgeries, still remain cyanosed.

Table 2:

Complications of Congenital heart disease

1. Left or right ventricular outflow tract conduit / homograft stenosis or incompetence.
2. Post stenotic left or right ventricular outflow tract aneurysmal dilatation or dissection.
3. Intractable arrhythmias and resultant cardiomyopathic ventricles.
4. Patients may have sequential pacemakers: older models may need to be reset prior to surgery to prevent diathermy interference.

5. Poor ventricular function from volume or pressure overload.
6. Atrio-ventricular valve regurgitation or stenosis.
7. Irreversible pulmonary hypertension.
8. Multiple arterial pulmonary collateral arteries (MAPCAS) resulting in systemic arterial desaturation.
9. Protein losing enteropathy from longstanding cardiac failure
10. Associated with this: life threatening plastic bronchitis in the lungs from proteinaceous casts associated with pulmonary venous hypertension.
11. Systemic hypertension
12. Co-existent Ischaemic Heart Disease with long-standing CHD
13. Congestive cardiac failure from burnt out CHD with or without embolic phenomena.

Any patient with associated exercise effort intolerance should be investigated to rule out deterioration in CHD or the presence of associated overt ischemic heart disease. Signs of poor cardiac function may be very subtle, patients with CHD may be used to long standing effort intolerance and have been able to adapt physiologically for many years, chest pain may not always be ascribed to angina, therefore these patients need to be investigated very carefully prior to elective incidental surgery. Congestive Cardiac failure with CHD is receiving more attention. Anti-failure therapy, B-blockade, afterload reduction is more commonly prescribed and may impact on anaesthesia. (6)

The complication of pulmonary hypertension either as a primary diagnosis or secondary to CHD is a very significant risk factor for pregnancy. A recent review found that despite modern trends in the use of pulmonary vasodilator agents, maternal mortality associated with either regional or general anaesthesia, still carries a 36% mortality incidence. (7) These reviewers consider that discouraging pregnancy in patients with significant pulmonary hypertension is the wise management choice to protect the patients life (7).

Specific anatomical considerations are important particularly for more complex CHD. This depends on the age of the patient and the era in which the cardiac surgical repair was completed. For example; the patient may previously have undergone surgical repair for complex congenital heart disease such as transposition of the great vessels with an atrial switch procedure the;(Mustard or Senning) operation. In this case attention directed at detecting baffle leak or baffle obstruction may well be required. Medical control of arrhythmias or radiofrequency arrhythmia ablation may be needed as these patients are particularly prone to supra-ventricular arrhythmias because of long intracardiac surgical suture lines. These patients after 15 to 20 years invariable may have a pacemaker that would need to be checked for adequate function prior to incidental surgery. These last two cardiac operations (Mustard and Senning) have for the most part been replaced with the arterial switch procedure in latter years if the anatomy is favorable. Currently these patients do very well, although coronary artery stenosis remains their most significant problem.

Specific considerations for Tetralogy of Fallot include markers for the risk of arrhythmias and sudden death (Table3).

RVSP > 60 mmHg

RV-to-PA gradient > 40 mmHg

Repair at >12 age, not protective

Ventriculotomy not protective

Co existing aberrant LAD origin 3-5%

QRS duration > 180 ms = 100% Ventricular Tachycardia risk

QRS duration increasing at >greater than 3.5 ms/yr

Adapted from: Steeds RP QJ Med :7-13 1997 ref (8)

Secondly, if palliation for *single ventricle anatomy* was completed in the last 20-30 years, it is important to determine which operations have been done. A modified aorto-pulmonary (Blalock Tausig) shunt (MBTS) to increase blood flow to the lungs in order for the pulmonary arteries to grow, may have been done during early infancy. This procedure is usually completed on the right side, in which case importantly for anaesthesia, blood pressure measurement in the right arm will under read.

Sometimes when blood flow to the lungs is excessive with single ventricle physiology during infancy, a pulmonary artery banding procedure is completed to protect the lung vasculature from developing pulmonary hypertension. Either of these procedures (Shunt or Band) is usually then followed by a bidirectional cavopulmonary anastomosis or (Glenn) operation in which the superior vena cava is anastomosed to the pulmonary artery. This operation is usually completed during infancy prior to the modified total cavopulmonary anastomosis or (Fontan) where the inferior vena cava is then also

anastomosed to the pulmonary artery. The Fontan operation is usually completed during the toddler years, however it can be completed as an adolescent or adult if the pulmonary vasculature has been protected from hypertension and systemic ventricular function has been preserved. Following the Fontan, patient arterial saturation usually returns close to 100 %. This modified Fontan completion means all venous return effectively goes to the lungs passively and is dependent on the adequate functioning of the cardiorespiratory pump interaction. Here, spontaneous ventilation ultimately helps venous return to the lungs for oxygenation and return to the left atrium to ensure adequate cardiac output. If this procedure has been completed with a fenestration from IVC to common atrium, for fear of the patient developing intractable ascites, or protein losing enteropathy, then, patients may still be marginally desaturated with pulse oximeter readings of nearly 90%.

The point of this sequence of three cardiac operations usually well before adolescence, is that the initial congenital heart disease diagnosis was of such a severe nature that a two ventricular repair was never going to be possible. This management decision is usually taken within the first few months of life. Patients with single ventricle anatomy do not have a second ventricle to pump blood to the lungs and all adequate lung blood flow is subsequently dependent on the transpulmonary pressure gradient. By adulthood these patients may have developed valve stenosis, regurgitation, aortic arch stenosis, arrhythmias and significant ventricular dysfunction.

Usually this series of three operations resulting in passive pulmonary blood flow determines the anesthetic management principles in the patient with Fontan type

physiology (Figure 1). This depends on ensuring an adequate transpulmonary arterial-to-venous pressure gradient. If SVC or IVC obstruction occurs, passive pulmonary blood flow will be limited. If the pulmonary venous pressure is high because of a high LVEDP, AV valve stenosis or regurgitation, or loss of sinus rhythm, the pressure gradient decreases and there will be poor forward blood flow through the lungs. Low cardiac output associated with hypoxia will result. Spontaneous ventilation preserving cardio-respiratory interactions is preferred, but if intubation and ventilation are required for anaesthesia and surgery, then ventilation with peak airway pressure below 30cm H₂O and mean airway pressure less than 10 cmH₂O with a long expiratory time, optimizing passive pulmonary blood flow is recommended. An adequate preload is required and SVR should be maintained to optimize coronary perfusion. These patients are at risk for perioperative thrombosis due to low flow and venous stasis of blood. It is important perioperatively to check if anticoagulants are part of the medical regimen as this could preclude the use of central neural blockade because of the risk of epidural or subarachnoid hematoma if regionals are used as part of the anaesthetic technique. If any baffle leaks are present, intravenous air-filters are recommended as part of the anesthetic management to lower the risk of air emboli being showered to the arterial system.

These three stepwise palliative cardiac operations may be undertaken for multiple cardiac diagnoses and each of the surgeries creates quite different CVS anatomy and although seen predominantly in children, some adolescents may need to have conduit revision surgeries or valve replacements. These patients also present for non cardiac surgery and each diagnosis requires a specific anaesthetic plan.

Thirdly, establish preoperatively if the patient is of adequate nutritional status and that protein losing enteropathy is not present. Will they be fit enough nutritionally to recover from major surgery? Will a period of recovery in a cardiac intensive care unit be required? Does the patient have a diagnosis of any specific syndrome and has there been adequate optimization of co-morbidities: including hypertension, renal disease, ischemic heart disease, epilepsy, blood, liver, or respiratory disease (1).

Particularly in the adult patient with surgically corrected CHD that now has a structurally normal heart but poor myocardial function, is there any evidence of coronary artery disease? The diagnosis of coronary artery disease is often overlooked in this patient population group and cardiac symptoms from ischemic heart disease that progress, may wrongly be attributed to deteriorating CHD.

Establish exercise effort tolerance, CVS medications, and time of last cardiological examination (preferably within the last 6 – 12 months), gathering data from the recent echo or cardiac catheterization. For elective surgery, the patient should be free of any current URT or pulmonary infection and will need infective endocarditis prophylaxis for surgery.

Finally, in order to plan the anaesthetic technique, we specifically want to know more about: *Current preserved myocardial function*: What is the cardiac ejection fraction? Is any valvular regurgitation, stenosis or myocardial arrhythmia present? Are

there any residual intra-cardiac anatomical defects creating L-to-R or R-to-L shunts? What is the magnitude of these shunts as defined by the Qp:Qs ratio, could the shunt be contributing to LV or RV volume overload? Have any stents, occluder devices or balloon dilatations been required to correct residual stenotic defects to preserve LV or RV function? Has a low pulmonary vascular resistance been preserved preventing RV pressure overload? Has the patient had a myocardial infarct, or is there any evidence of reversible myocardial ischemia from coronary artery insufficiency that may require cardiological angioplasty and stenting or coronary artery bypass grafting prior to elective major abdominal surgery?

Once all this information is known, the patient medically optimized and the complexity of the planned surgery has been established, the degree of intraoperative invasive monitoring, anaesthetic technique and postoperative recovery plan are then decided upon. Intraoperatively, do not use excessive PEEP in patients with a total cavopulmonary anastomosis. In this group of patients avoid the trendelenberg position for any protracted period of time; they do not tolerate the cardiovascular changes well.

Monitoring

Standard EKG, blood pressure cuff, pulse oximetry, agent, oxygen analyzer, capnography are mandatory. It may be necessary to place an indwelling arterial catheter under local anesthesia prior to induction of anesthesia in patients with poor myocardial reserve. There are indications to use an internal jugular central venous catheter for drug

delivery and optimization of preload in ill patients undergoing extensive surgery. The transesophageal echo is a valuable monitor and diagnostic tool able to differentiate a myocardial anatomical defect from a myocardial functional problem in the presence of low cardiac output states intraoperatively. It is also a very valuable diagnostic tool detecting early myocardial ischemia when regional wall motion abnormalities are noted.

Induction of anesthesia

Importantly the myocardial substrate may not be normal. Recently intramyocardial calcification has been described utilizing MRI. The cause of this calcification may be from the initial surgery with myocardial protection problems, or ongoing myocardial fibrosis and calcification over time. Specifically, care is needed with the dose-dependent potent myocardial depressant effects of inhalational anaesthetic agents in patients with poor myocardial function. Most children and adolescents with CHD will tolerate a careful inhalational induction. Sevoflurane exhibits a more stable hemodynamic induction and maintenance profile than halothane for cardiac anaesthesia (9). Adults with CHD and severe myocardial dysfunction may tolerate an intravenous induction and endotracheal intubation (with etomidate or fentanyl, midazolam, muscle relaxant) better than an inhalational agent. Etomidate in atrial and ventricular tissue studies from failing hearts, has been shown to have no myocardial depressant effects when used at clinical doses (10). Any patient with hypertrophic cardiomyopathy, critical aortic stenosis, adult uncorrected Tetralogy of Fallot or Eisenmenger's Syndrome is not a candidate for a propofol anesthetic induction. The reason is a 20-30% drop in systemic

vascular resistance with this agent may decrease coronary perfusion or significantly decrease the Qp:Qs ratio across an intracardiac shunt, causing hypoxia. Figure 2 (11)

Inspired Oxygen

The role of low FiO₂ and maintaining low oxygen saturation is seldom required in adults with CHD since the surgical stages of palliation should have been completed in childhood. The one time it may be appropriate to utilize low FiO₂ during anesthesia for adults with CHD, is in the cardiac catheterization laboratory where an adult patient with end-stage CHD may be undergoing a diagnostic heart cath to determine if the pulmonary vascular resistance is low and then reactive when 100% oxygen is administered. This would be done prior to being listed as a candidate for heart transplantation. Low FiO₂ and oxygen saturation of 80 % may be appropriate only for single ventricle neonates pre and post surgery in the first stage of single ventricle palliation surgical procedures and importantly, only use a low FiO₂ if Cardiac Output is maintained.

Adult patients with severe CHD may manifest cyanosis. This is a particularly worrying clinical sign and investigations including echocardiography and possibly cardiac catheterization prior to elective non cardiac surgery are often required. It is possible that the patient may have an intracardiac baffle leak, VSD or ASD repair leak that may be amenable to repair with an intracardiac occluder device prior to surgery. This could be done in elective cases to decrease the risk of right to left embolic events that may lead to cerebral embolic events. Hypoxia induced polycythemia limits maximal

exercise and together with dehydration predisposes these patients to vascular thrombosis particularly cerebral (4).

A more sinister diagnosis (although unusual in TAPVC) of cyanosis occurs in patients with non reactive pulmonary hypertension. This diagnosis identifies a patient at extremely high risk for any type of anesthesia. Non reactive pulmonary hypertension in surgically corrected CHD is invariably associated with severe right ventricular pressure overload, right ventricular hypertrophy and dilatation, leading to right ventricular dysfunction, a pulmonary valve incompetence murmur, tricuspid regurgitation and evidence of ascites or pedal edema are often present. If this diagnosis is due to long standing uncorrected CHD, sometimes from a simple unrestrictive ASD or VSD causing prolonged left to right blood flow and the development of fixed non reactive pulmonary hypertension; this is usually referred to as Eisenmenger's syndrome. Regional anesthesia procedures are preferred in patients with this diagnosis if possible.

The anaesthetic aims in patients with pulmonary hypertension are to avoid: acidosis, alveolar hypoxia, hypercapnia, atelectasis and hyperinflation of the lungs and prevent high airway pressures (4). These hemodynamically critically balanced patients require invasive monitoring for all but very simple short duration surgeries and should recover in an ICU and seldom be done as outpatients. General anaesthesia should not be undertaken lightly without consideration of all the associated risks described above.(12)

Summary

The management of a grown up patient with CHD depends on the age of the patient and the surgeries completed. Remember that the last cardiac surgical procedure may well have been a heart transplant for burnt out CHD. If so, special considerations for excluding accelerated obliterative coronary artery disease needs to be considered. The heart transplant patient may also have silent myocardial ischemia, chronic rejection and ventricular dysfunction, myeloproliferative disease and may be immunosuppressed.

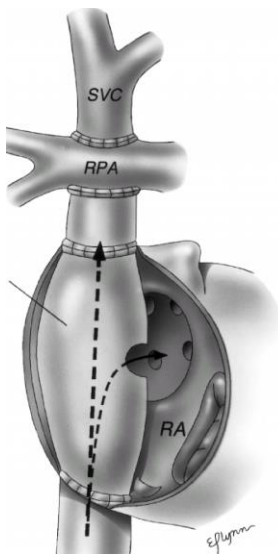
Perioperative anaesthetic checklist

1. Simple or complex CHD?
2. Cyanotic or acyanotic cardiac lesion?
3. Exercise effort tolerance? A very important feature of adequate cardiac function.
4. Single ventricle anatomy or two ventricle repair?
5. Absence or presence of pulmonary hypertension; If present is pulmonary hypertension reactive or non reactive? If non reactive life threatening surgery only
6. Preservation of cardiac valve competency or presence of myocardial dysfunction?
7. Arrhythmias present: Medically treated or radiofrequency ablation?
8. Total cavopulmonary anastomosis or Fontan Physiology? Optimize ventilation, preload and consider inotropic support for myocardial dysfunction. A period of post operative ICU convalescence.
9. Has the patient had a heart transplant as one of the surgical procedures?
10. Optimize co-morbidities and other end organ damage.
11. Medications, including digoxin levels, electrolytes, anticoagulants?

12. Infective endocarditis (IE) present ? Document end organ damage
13. Ensure perioperative infective endocarditis prophylaxis.
14. Plan intraoperative monitoring, induction, maintenance and post operative care.

Figure 1

Fontan Physiology showing a fenestrated conduit. Fenestration preserves 15-20% CO.



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