The Adult with Congenital Heart Disease & Non Cardiac Surgery

Introduction
Adults with congenital heart disease are an increasing population with challenging physiologic considerations that are often in conflict with the operative techniques. Not surprisingly these scenarios are presented on fellowship and Board exams. The audience here at the SCA is well aware of this expanding population and the considerations for cardiac surgery. The challenges for the next decade are the increasing complexity of these patients and the organization of their care for elective, urgent and emergent non cardiac procedures. The number of adults living past 18 years will mandate that many of these procedures will not occur in specialized tertiary care centers and patients will want to explore care in their immediate community. The median age of adults with severe ACHD in the United States in 1985 was 17 and in the year 2000 it was 29 years. As cardiac anaesthesiologists, echocardiographers and ICU physicians we have a special understanding of their unique physiology and can play a leadership role providing directly or advising colleagues in the community how best to provide care for these patients. This expertise is not limited to the operating room but also in the pre-operative optimization and planning as well post operative monitoring and pain management. In conjunction with our ACHD cardiology colleagues we must be our own “Outreach Team” for this special population and outline physiologic parameters and goals for the post op recovery.

In this review of ACHD for non cardiac surgery we will discuss general considerations for patients with simple unrestricted shunts, palliated patients with surgical shunts, cyanotic heart disease, pulmonary hypertension and the patient that has had multiple interventions and/or coexisting syndromes. Many ACHD patients are dependent on a precarious balance of the PVR and SVR or are dependent on a low PVR to fill the left side and frequently will be considered for cases that require insufflations of carbon dioxide into a body cavity or ventilation strategies, such as a rigid bronchoscopy, that will adversely affect PVR. Respiratory, haematological and psychosocial issues are also major considerations with ACHD patients.

Up to and over 50% of patients with ACHD are lost to follow up in the transition from a paediatric institution to an adult setting. Ideally we should have comprehensive information about the patient’s anatomy and hemodynamics but we are often presented with emergent non-cardiac surgery and only basic information such as “He has tetralogy of Fallot repaired in childhood” and no access to previous records. We will review the complex lesions of Ebstein’s anomaly, Tetralogy of Fallot, Transposition of the Great Arteries and Single Ventricle patients palliated with the Glenn and Fontan procedures and the anaesthesia considerations for these particular types of ACHD lesions.

We will discuss several cases of non cardiac procedures in complex ACHD patients and review the anaesthetic considerations for this difficult group of patients.
General Considerations

Cardiac anaesthesiologists are accustomed to the associated defects in ACHD patient with syndromes such as "Catch 22" in Tetralogy of Fallot, VACTERAL syndrome and the cardiac defects of Down’s syndrome. The ACHD patient population has diminished static lung function and a reduced capacity to augment cardiac output in response to stress. Although unlikely to affect the ability to ventilate the patient intra-operatively diminished pulmonary function will have consequence in the recovery room or ICU and the ability to wean from the ventilator. Patients with cyanotic heart disease have erythrocytosis, an increase in the red cell line in response to hypoxia, and relative to the red cells a deficiency in vitamin K dependant clotting factors and platelets. Ironically these patients at an increased risk of thrombosis yet are frequently coagulopathic intra-operatively. Phlebotomy is inappropriate unless there are extreme symptoms of hyperviscosity as iron deficiency is an independent risk factor for stroke and thrombosis. Iron deficient red cells are less flexible in the microcirculation. Impaired renal function, as in patients with acquired heart disease is a significant predictor of morbidity and mortality. 1102 adult patients attending the Royal Brompton over a period between 1999 and 2006 had a serum creatinine and a glomerulo-filtration rate calculated. Renal dysfunction was mild in 41% of patients and moderate or severe in 9%. Not surprisingly severe renal impairment correlated with increased mortality over the 4 year follow up period.

Patients with simple unrestricted shunts such as ASDs, VSDs, large PDAs, and Aortopulmonary collaterals will have a left to right shunt in the presence of normal or low pulmonary vascular resistance and pulmonary blood flow may exceed systemic blood flow. A recently created Australian National registry for PAH in ACHD found that, of the first 50 registrants, 47 had systemic artery to pulmonary artery shunts. At rest breathing room air these patients may have a 2:1 or as much as a 3:1 shunt ratio of pulmonary to systemic blood flow. A common error is that an anaesthesiologist being concerned about the patient’s heart disease will ventilate the patient on 100% oxygen and unintentionally lower the patients PVR converting a 2:1 shunt to a much more severe imbalance such as a 4-5:1 shunt and ironically the pink patient will have a metabolic acidosis from diminished systemic oxygen delivery. Unrestricted left to right shunts have the potential to reverse with profound drop in the SVR. A well intentioned attempt to avoid a GA in a patient with a common atrium by using a spinal may trigger profound hypoxemia that may be difficult to reverse. Cyanotic heart disease patients with single ventricles and pulmonary circulation supplied by Aortopulmonary collaterals have a precarious balance between pulmonary blood flow and systemic oxygen delivery. After induction the anaesthesiologist should try to maintain the patient on a FiO2 that recreates there room air saturations at rest. Ventilation should be at lung volumes that are close to FRC and maintain a normal PaCO2.

Pulmonary hypertension is a major peri-operative risk factor. Eisenmenger’s syndrome is a particular challenge as the anaesthesiologist must deal with both the pulmonary hypertension and the potential to exacerbate the right to left shunt and cyanosis. Mayo clinic has reported a series of ACHD patients with Eisenmenger’s syndrome that had non-cardiac surgery in both community and teaching hospital settings. There was a death in each of 2 similar sized groups and no conclusion could be made about the safety of surgery in a nontertiary center. There is an exhaustive three part coauthored by 6 directors of
ACHD centres in North America, Europe and Japan with management recommendations.\textsuperscript{xv}\textsuperscript{xxvi} Challenging non-cardiac surgery cases will be presented.

Laparoscopic and video assisted surgery is increasingly becoming the standard for patients in general, gynaecological, urological and thoracic operations. Although insufflation of CO2 is in conflict with the physiologic goals intra-operatively in ACHD patients it bodes well for their recovery post operatively. Patient positioning (Prone, lateral, trandellenburg or head up) may exacerbate or alleviate this conflict. A clear plan with threshold for conversion to an open procedure must be discussed ahead of time with the attending cardiologist and surgical team as they may not be under our direct supervision. Although more challenging for the anaesthesiologist the patient will benefit from a closed procedure and avoid the complications of a large incision. Intraoperative monitoring of arterial and right atrial (caval) filling pressures with frequent sampling of arterial gases will be necessary when contemplating a laparoscopic approach.

Rigid bronchospy for pulmonary haemorrhage or suspension laryngoscopies for glottic and subglottic complications of prolonged intubation are cases not unusual in ACHD patients. Unlike the above discussion of laparoscopic vs. open the surgeon and the anaesthesiologists do not have much choice about technique and it is not the choice of anaesthesia agents but the maintenance of physiologic goals that is the challenge.

Pacing and defibrillator pads are best applied before prepping, draping and positioning with a defibrillator attached or in close proximity. Infective endocarditis guidelines have lessened but indicated in patients with prosthetic material, palliative shunts or conduits within the 1st 6 months of surgery or implantation but remain indicated if there is a residual lesion in close proximity to the repair. \textsuperscript{xviii} Example and AI jet hitting a VSD patch in a repaired tetralogy.

Epidurals both lumbar and thoracic can be used for open procedures however the anaesthesiology team will have to be involved in post operative monitoring and guidelines for the nursing staff. Clear blood pressure, central line pressures and expected oxygen saturations should be charted and a clear means of communication established for post operative recovery. Intravenous PCA is also a concern as narcotics will inhibit respiratory drive and make the PVR dependant patients vulnerable to rising PaCO2.

**Ebstein’s**

Ebsteins’ anomaly is a rare ACHD defect, less than 1% of all ACHD, that may present in childhood or as an adult with severe tricuspid regurgitation due to apical displacement of the septal and posterior leaflets of the TV and a small “atrialized” thin walled RV that is hypo kinetic. The patient may shunt right to left across a PFO or ASD; these patients require an adequate filling pressure and a technique that minimizes the PVR. \textsuperscript{xxvii} They patient will often be unaware of their diagnosis and present for cardioversion of new onset atrial flutter.
Tetralogy of Fallot

Tetralogy of Fallot rarely presents in the adult world uncorrected. Older adults likely will have had a temporizing systemic artery to pulmonary artery shunt, Blalock Taussig, Potts or Waterston Coley, and a definitive repair as an older child. Younger adults are likely diagnosed in utero and have had a primary correction as a neonate. Most patients will have had a transannular patch to widen the RVOT at the time of repair and over time they will develop pulmonary insufficiency, right ventricular dilatation and eventually tricuspid regurgitation due to right ventricular dilatation. Tetralogy patients are susceptible to ventricular tachycardia, atrial arrhythmias and sudden death and a widened QRS complex >180 ms correlates with an increased risk of sudden death. Unfortunately asymptomatic Holter monitor detected V tach does not correlate with sudden death and AICDs in these patients has not been shown to diminish the risk of sudden death. Like the Ebstein’s patients with repaired tetralogy require an adequate filling pressure and a technique that minimizes the PVR.

Transposition of the Great Arteries

D-TGA with a Mustard or Senning Baffle

D-TGA patients have atrial-ventricular concordance and ventricular-great artery discordance. The RV is the sub aortic and systemic ventricle. Although rarely performed now there are many adults living with atrial baffle repairs which re-directed the atrial blood to the opposite ventricle. The RV remains as the systemic ventricle. Patients with a Mustard or Senning repair frequently have baffle leaks, sinus atrial arrhythmias, sick sinus syndrome and inevitably systemic ventricular failure. Although their anatomy may be confusing they are managed much like a dilated cardiomyopathy waiting for a heart transplant with an AICD or PPM in-situ.

D-TGA with an Arterial Switch

Early in the 1980s CHD paediatric centers moved from the above repair to the Jatene or Arterial Switch operation. The aortic and pulmonary arteries are switched with re-implantation of the coronary arteries. These patients have yet to reach adult centers in large numbers but complications include coronary artery perfusion defects and aneurysms of the neo-aortic root. The advantage is that the LV is the systemic ventricle AV and VA synchrony is restored. Urgent non-cardiac surgery can be done safely in the community setting with consultation from cardiac anaesthesia and an ACHD cardiologist.

D-TGGA with VSD and Pulmonary Stenosis and a Rastelli Repair

Patients with D-TGA (AV concordance and VA discordance) having a VSD and Pulmonary stenosis are corrected in childhood with a Rastelli repair which entails a tunnel from the LV through the VSD to the aorta and an extra cardiac conduit that connects the RV to the main PA. A red cell flowing through the heart will travel through all the chambers in an appropriate sequence and the LV is restored as the systemic ventricle. Frequently the RV to PA conduit will become obstructed and require balloon dilatation or replacement. Anaesthetic management must account for stenosis of the conduit and occasionally patients will have sub aortic obstruction.
**L-TGA**

L-TGA is also called congenitally corrected transposition which is a term that confuses many community and non cardiac anaesthesiologists. The term implies that the patients are “born corrected”. These patients have both AV discordance and VA discordance. The RV is RA is connected to a morphological LV that is subpulmonic. The LA is connected to a morphological RV which is sub aortic. The majority of these patients have an Ebstein like malformation of the tricuspid, systemic AV valve, with moderate to severe systemic AV valve regurgitation. In the absence of associated anomalies these patients present in the 2\(^\text{nd}\) and 3\(^\text{rd}\) decade of life with AV valve regurgitation and symptoms of heart failure. They can also present with heart block. Much like the Mustard patients they are managed much like that of a dilated cardiomyopathy.

**Single Ventricle with Glenn and Fontan Type Connections (Tricuspid Atresia and Pulmonary Atresia)**

Patients with single ventricle physiology such as tricuspid atresia, pulmonary atresia and DILV will have had likely have had a Glenn shunt followed by a form of the Fontan procedure. A classic Glenn is a SVC directly anastomosed to the RPA; a bi-directional Glenn is the SVC anastomosed to the confluence of the LPA and RPA. The Fontan palliation was first described in 1971. Originally the operation was an anastomosis of the RA to main PA and subsequent modifications have included the lateral tunnel Fontan and the total Cavopulmonary connection. These patients have no ‘pump’ for systemic venous return to get to the left atrium. For successful management they must have adequate right sided filling pressures, low PVR, unobstructed pulmonary vein inflow to the LA, AV synchrony, a competent systemic AV valve with laminar inflow, no outflow tract obstruction and an afterload reduction for the systemic ventricle. Patients may have had a fenestration from the Fontan connection to the systemic or common atrium and precautions for venous emboli to the systemic circulation will be needed. Anaesthetic techniques must be chosen that keep all of these goals in mind but also the planning of the post operative recovery and step down must take them into account. Spontaneous respiration is desirable but not always compatible with the surgical plan. As anaesthesiologists we may accept increase risk in our monitored environment for improved hemodynamics in the post recovery phase. xiii

**Summary**

Management of the ACHD patient is challenging and rewarding. Many patients will be receiving ongoing monitoring an support however in an established ACHD center but frequently we will be the patient’s first contact with a specialist physician in several years of having been lost to follow up. We are in a unique situation to identify a patient at risk and in addition to pre, intra- and post operative care reconnect them to a an ACHD team for subsequent care.