Adult Congenital Heart Disease Review

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Background and Scope

It is known that we start with a prevalence of congenital heart disease (CHD) in children of between 0.5 and 1% of live births of which approximately 1.5 per thousand have complex CHD. With interventional treatment, more than 90% of the children with CHD will now survive into adulthood compared to only 15-25% survival for untreated patients. In the ACHD population, nearly 50% of patients require ongoing follow-up and further procedural treatment. This matches a predicted need for follow-up in ACHD of greater than 200 per 100,000 live births. In 2006, it was estimated that there were approximately 125,000 ACHD patients in Canada. During the lifetime of ACHD patients who have had a previous surgical or interventional cardiac catheterization corrective or palliative procedure, half will eventually undergo two or more procedures and one quarter will have three or more procedures. The profile of this group has changed and can be expected to continue to change due to factors such as improved survival of patients with complex CHD, definitive repair or complex palliative procedures being undertaken at a younger age, as well as improvements in the procedures as well as the spectrum of care before, during, and after these procedures. One example is the increasing number and variety of complex single ventricular patients surviving through to completion Fontan. Another example is that the arterial switch operation has resulted in a group of patients with a nearly normal life expectancy in contrast to the group who underwent Mustard/Senning repairs and who were plagued by the late complications of systemic ventricular failure and dysrhythmias. It is also hoped that there will be fewer adult patients with end-stage pulmonary vascular disease and Eisenmenger's syndrome. Approximately 10% of the ACHD population are not diagnosed until adulthood. Common lesions in this group include Ebstein's anomaly, secundum ASD, and congenitally corrected TGA. Lastly, there is the changing and unpredictable impact of immigrants with ACHD arriving from countries where they may have had no access to cardiac surgery.

Some Specific Lesions

Atrial Septal Defect (ASD)

Comprising 7% of all CHD, ASDs make up approximately 30% of ACHD. Three quarters are ostium secundum defects occurring at the level of the foramen ovale. In untreated patients, mortality increases by 6% per year following the age of 40. The remaining types of ASDs include ostium primum endocardial cushion defects (15%), sinus venosus defects (10%) and rare cases of coronary sinuses draining into the left atrium (LA) (unroofed coronary sinus). Volume overload from the shunt from left-to-right atria results in dilation of the right atrium (RA) and ventricle, right ventricular (RV) hypertrophy and enlargement of the pulmonary artery (PA). Patients may be asymptomatic or may progress to develop shortness of breath, supraventricular tachycardia, and right ventricular failure. In severe cases tricuspid regurgitation may result from RV dilation. Eisenmenger's syndrome is rare

Sinus Venosus ASDs are associated with partial anomalous pulmonary venous drainage (PAPVD) in which one or two pulmonary veins drain into the systemic venous return. Most frequently the right upper pulmonary vein drains into either the base of the SVC or into the right atrium. These patients have right atrial and ventricular volume overload. Preoperative workups may miss one or both of the veins constituting the PAPVD.

In providing anesthesia for patients with an ASD, due to the magnitude of the circulating blood volume required to serve the shunt, hypovolemia is not well tolerated. The systemic vasodilation associated with potent inhalation anesthetics may diminish L→R shunting. PEEP and high ventilator pressures may lead to shunt reversal and the risk of paradoxical venous emboli. Intraoperative transesophageal echocardiography is helpful to monitor for left-sided air during and following ASD closure. Increased vigilance is also required when ASD closure is conducted under electrically-induced fibrillation in order to avoid the consequences of a disconnection from the fibrillator and a possible return to sinus rhythm whilst there is air in the left atrium that could lead to systemic air embolism.

Endocardial Cushion Defects

With a common embryological origin, endocardial cushion defects result in an ostium primum ASD with or without an inlet VSD (complete atrioventricular canal form) and have a common AV valve with clefts in the septal (most commonly mitral portion) leaflets. Volume overload from the left to right shunt results in dilation of the RA, RV, and left ventricle (LV) as well as biventricular hypertrophy. These defects are associated with Down syndrome and require early surgical intervention as pulmonary vascular disease can develop rapidly. Unrepaired complete forms rarely present in adults. However, accounting for approximately one tenth of ACHD surgical cases, adults with previously repaired endocardial cushion defects present for repair of progressive mitral regurgitation, subaortic stenosis, and pacemaker revision for atrioventricular (AV) block.

Patent Foramen Ovale (PFO)

PFO carries a risk of systemic air emboli with decompression following deep-sea diving as well as with laparoscopic procedures or sitting position surgery. The diagnosis may be confirmed by performing an echocardiographic contrast study in time with a valsalva maneuvre. PFOs have a population incidence of between 5 and 25 percent, and are typically only closed in conjunction with cardiac surgeries that require atriotomy.

Ebstein's Anomaly

In Ebstein's anomaly, accounting for approximately 0.5% of CHD, the inlet portion of the RV is atrialized and the RV cavity size is reduced due to the insertion of the septal and sometimes also the posterior leaflets of the tricuspid valve being displaced apically by more than 10 mm. Typically the anterior leaflet is large and dysplastic and short, tethered or fibrosed chordae tendonae impair leaflet motion. This may result in tricuspid regurgitation, stenosis, or both. One third of patients will have dysrhythmias including Wolff-Parkinson-White (WPW) syndrome (10%) or AV block. Prognosis is influenced by the size of the heart chambers, the patient's functional status (particularly the presence or absence of right heart failure), the degree of rhythm problems, and the presence of cyanosis which may occur when an ASD is associated with this lesion. In providing anesthesia for patients with Ebstein's anomaly, it is essential to account for their limited cardiac output and to optimize the impaired RV filling. Tachycardia is not well tolerated as it limits the diastolic filling time. Bradycardia is also not well tolerated. The massively dilated RA poses a significant threat of dysrhythmias during central venous line (CVL) insertion. Pulmonary artery catheters should be avoided. Milrinone is often selected in order to avoid inotrope-induced tachycardia. Tricuspid valve (TV) reconstruction may result in sufficient residual tricuspid regurgitation or stenosis to warrant TV replacement and AV block, if not already present may present following repair.

Ventricular Septal Defect (VSD)

Accounting for roughly 10% of ACHD VSDs are classified as 1) perimembranous or membranous (almost 70% of VSDs), 2) inlet type (accompany endocardial cushion defects), 3) infundibular or supracristal (often associated with prolapsed of the right coronary cusp leading to aortic regurgitation), and 4) muscular (most often multiple). Most of the volume of the left to right shunt bypasses the RV cavity due to the location of the VSD relative to the RV inlet and the RV outflow tract. Thus the LV is subject to increased pressure and volume work and there is enlargement of the LA and LV and dilation of the PA. RV hypertrophy only occurs following the onset of pulmonary hypertension (PHT). In ACHD, VSDs run a spectrum that includes: no or minimal residual shunt as a result of spontaneous or surgical closure, small and associated with a small shunt and normal pulmonary arterial pressure (PAP), a shunt large enough to result in elevated and reactive PAP and RV hypertrophy (RVH), and finally Eisenmenger syndrome (discussed in a subsequent section). The hemodynamic goals under anesthesia are to diminish the degree of L→R shunting. Potent inhalational anesthetics can assist this through lowering the systemic vascular resistance (SVR). In patients with large VSDs, the reflex systemic vasoconstriction that accompanies hypovolemia should be avoided as it may increase L→R shunting. Low FiO_2, positive end expiratory pressure (PEEP), and mild hypercarbia may be used to decrease the shunt by elevation of PVR in the absence of PHT. However, in patients with PHT the goal is to lower PAP and prevent increases in PAP (see also the subsequent section pertaining to PHT).

Tetralogy of Fallot (TOF)

Pediatric anesthesiologists are familiar with the lesions that comprise TOF: large membranous VSD, overriding aorta, right ventricular outflow tract (RVOT) obstruction (this often has a dynamic component, but may be accompanied by stenosis and thickening of the pulmonary valve (PV) and/or by hypoplasia of the pulmonary arteries), and RVH. A further 25% have an accompanying ASD (Pentalogy of Fallot) and there is a 10% incidence of anomalies of the coronary circulation.

TOF is the third most frequently encountered lesion in ACHD practice. In the absence of surgery, survival is estimated at 30% at ten years and below 3% at forty years. Survival into adulthood without surgery depends upon preserved pulmonary blood flow either due to the presence of mild RVOT obstruction or the development of multiple aortopulmonary (AP) collaterals. Complete repair in infancy has replaced palliation by means of Blalock-Taussig (BT) shunts followed by definitive childhood repair. Infant TOF repair resolves cyanosis, circumvents dysfunctional RV remodelling, and facilitates growth of the pulmonary vasculature. In contrast, BT shunts were complicated by ongoing cyanosis, compensatory erythrocytosis, and ventricular remodelling due to RV pressure overload and LV volume overload.

ACHD patients with prior TOF repair present to the surgical suite with long-term complications such as: dysrhythmias (right bundle branch block, bifasicular block, atrial fibrillation, atrial flutter, and PVCs in more than 50%), residual VSDs (10-20%), obstruction of an RV to PA conduit or the RVOT, pulmonary or conduit valve regurgitation or stenosis, RV dysfunction from volume overload, and tricuspid regurgitation. A thorough knowledge of each patient's combination of lesions is essential to anesthetic and hemodynamic management. Residual pulmonary regurgitation may be aided by careful mechanical hyperventilation to achieve hypocarbia and hyperoxia whilst limiting ventilator pressures. Maintenance of systemic arterial pressure and a normal heart rate are important in the management of the subendocardial RV ischemia that may accompany residual pulmonary stenosis.

In uncorrected patients, anesthetic management aims to maximize pulmonary blood flow and minimize the R→L shunt through: 1) avoidance of hypovolemia as dynamic outflow tract obstruction worsens in the under-filled ventricle and the associated hypotension will increase R→L shunting, 2) systemic arterial vasoconstriction (i.e., phenylephrine), and 3) relief of dynamic outflow track obstruction with judicious β-blockade. Pulse oximetry is the most useful monitor for tracking these changes.

It is crucial to ensure adequate pulmonary blood flow by maintaining systemic diastolic blood pressure in patients dependent upon BT shunts or AP collaterals who often have an already a low diastolic BP due to pulmonary run-off. As well, their chronically overloaded LVs are at risk of decompensation.

Dextro-Transposition of the Great Arteries (d-TGA)

VSD accompanies d-TGA in 30-40% of patients. The arterial switch operation (Jatene) is the mainstay of modern surgical treatment. It is associated with a low mortality rate and a predicted almost normal life expectancy. Rare complications include neoaortic regurgitation and or coronary ostial lesions. ACHD patients who underwent the atrial switch (Mustard or Senning) operation had a complex atrial baffle placed in order to direct systemic venous return to the subpulmonic ventricle and the pulmonary venous return to the subaortic ventricle. These patients may require reoperation due to baffle obstruction of the pulmonary or systemic venous return. These complicated atrial repairs also lead to difficult to treat atrial tachydysrhythmias and AV blocks in more than half of patients. Eventually, forcing the RV to perform systemic work leads to progressive RV dilation and dysfunction that result in failure after 20 to 25 years.

Levo-Transposition of the Great Arteries (Congenitally Corrected Transposition, l-TGA)

In l-TGA the aorta and PA as well as the RV and LV are transposed (atrioventricular discordance with ventriculoarterial discordance). This may be accompanied by a perimembranous VSD (80%), mitral anomalies (50%) and tricuspid anomalies (30%). As well, the misalignment of the interventricular septum and the great vessels may lead to pulmonary outflow track obstruction. The majority do not become symptomatic until adolescence. Atrioventricular block occurs commonly and the systemic ventricle dilates and fails after 20 to 25 years.

Bicuspid Aortic Valve (BAV)

Affecting 2% of the population, BAV is the most common congenital cardiac anomaly. Of disparate size, the two cusps are fragile and subject to stress from abnormal coaptation. Over the course of years, this leads to calcification and stenosis whilst associated annular dilation may produce aortic incompetence. BAV is associated with coarctation of the aorta and perimembranous VSD.

Coarctation of the Aorta

Coarcation of the aorta occurs in approximately 2 per thousand live births. Half of the patients with coarctation of the aorta will have a BAV. Pediatric anesthesiologists are familiar with the infant presentation of the more severe forms of coarctation following post-natal closure of the patent ductus arteriosus (PDA). However, one in five cases will have a persistently patent PDA. Pressure overload leads to LV hypertrophy (LVH) accompanied by diastolic dysfunction, but with preservation of systolic function. Collaterals (internal mammary, intercostals, cervical, and intercostals) develop in patients with severe coarctation. Without surgery, complications (i.e., LV failure, aortic rupture, intracranial hemorrhage, and endocarditis) lead to death in the fourth or fifth decade of life. Although coarctation of the aorta is commonly repaired in children without the use of lung isolation, the use of a double-lumen endotracheal tube may facilitate the left thoracotomy in adults. Activation of baroreceptors accustomed to the previous proximal hypertension contributes to early postoperative hypertension whilst peaking of rennin and angiotensin contributes to postoperative hypertension on the second to third postoperative day. Between 20 and 50 percent of patients will have persistent hypertension accompanied by LVH. Paraplegia from spinal ischemia is a rare operative complication (0.1 to 0.4%). Recurrence of coarctation is not uncommon.

Fontan Circulation

The Fontan procedure was first described for the palliation of tricuspid atresia in 1971. Currently bidirectional cavopulmonary anastamosis (BCPA), often referred to as bidirectional Glenn shunts, followed by completion Fontan repair are used in the palliation of a wide variety of complex single ventricular anomalies. Initially many of these infants will have first undergone a Norwood, Norwood-Sano, hybrid, BT shunt or pulmonary artery band procedure depending upon the lesion. Along with

improved multi-stage survival, this more diverse Fontan palliation cohort will lead to a different population of adult Fontan survivors. Another surgical change that will influence this population has been the move towards the use of an extracardiac conduit for the completion Fontan rather than an intracardiac repair. The intracardiac atrial baffle repairs were complicated by RA dilation with subsequent blood stasis, thrombosis, and arrhythmias. Success of the Fontan depends upon: adequate pulmonary vascular bed, AV valve competency, good systemic ventricular function (anatomical left ventricles do better), and sinus rhythm. As many as 28% of patients with Fontan circulation may develop chronic systemic venous hypertension and stasis followed by ascites, hepatic dysfunction, coagulation disorders, and protein-losing enteropathy.

Pulmonary perfusion is driven by the CVP to left atrial pressure (LAP) gradient of approximately 8 to 10 mmHg (LAP of 5-8 mmHg and CVP of 13 to 18 mmHg). Not surprisingly, increases in PVR (hypoxia, atelectasis, hypercarbia, and acidosis) or increased intrathoracic pressure (positive pressure ventilation, valsalva maneuvres, PEEP) are not well tolerated. Elevated LAP as a consequence of AV valve regurgitation or systemic ventricular dysfunction is also poorly tolerated. Intraoperatively, inotropic support may be required. The loss of sinus rhythm associated with the dysrhythmias that may occur in up to 40% of patients with intracardiac Fontan completion are poorly tolerated due to diminished filling of the systemic ventricle and elevation of the LAP.

Anesthetic care requires fastidious attention to factors that maintain pulmonary blood flow (PBF). Hypovolemia can lead to a catastrophic reduction in PBF. Despite the importance of maintaining CVP, central venous lines are used very selectively due to the risk of thrombosis. The rise in intrathoracic pressure during inspiration interrupts pulmonary blood flow. Therefore if positive pressure ventilation is required the strategy should be one that avoids PEEP and uses low inspiratory pressures to achieve mild hypocarbia. A postoperative return to spontaneous ventilation as soon as possible is important as inspiratory subatmospheric intrathoracic pressure helps drive pulmonary blood flow by aspirating blood into the PA.

Last but not least many of these patients are on prophylactic anticoagulation and will need a plan for their perioperative coagulation management and the postoperative resumption of anticoagulant therapy.

In patients with Fontan circulation undergoing non-cardiac surgery, epidurals may be used cautiously whilst carefully maintaining the preload. Spinal anesthesia is ill-advised as the drop in preload occurs too rapidly and the possibility of epidural hematoma is increased due to venous congestion and anticoagulants administration.

Eisenmenger Syndrome

Representing the end-stage of patients with PHT as consequence of long-standing untreated left to right intracardiac or extracardiac shunts (see also the subsequent section on PHT). The features of Eisenmenger syndrome include severe, fixed pulmonary hypertension with equalization of the RV and LV pressures, bidirectional or even reversed (R→L) shunting and eventual dilation and failure of the RV accompanied by tricuspid regurgitation. Forty percent of patients will die prior to the age of 25 years with a median survival in the mid fourth decade. Predictors of mortality include: syncope, increased age at onset of symptoms or diagnosis, supraventricular dysrhythmias, poor functional class, severe RV dysfunction, elevated RA pressure, SpO_2 below 85%, renal insufficiency and Down syndrome. Pulmonary vascular resistance (PVR) may exceed 800 dynes/s/cm^5 and a ratio of PVR to SVR greater than 0.7 signifies prohibitive surgical risk.

In all but the simplest of cases, an arterial line is essential monitoring in order to achieve the hemodynamic goal under anesthesia of preserving the balance between SVR and PVR. As the PVR is fixed, the goal is to avoid decreases in SVR. Hence volatile-agent-induced vasodilation may be problematic. In non-cardiac surgery neuraxial anesthesia is poorly tolerated, but when feasible regional nerve blocks can be a good solution. Sudden increases in PVR must be avoided as 1) in patients with intracardiac shunts this may trigger oxygen desaturation followed quickly by a fall in cardiac output, bradycardia and cardiac arrest or 2) in patients with extracardiac shunts acute RV failure may produce the same rapid decompensation. Bearing in mind its potential to increase myocardial oxygen consumption, ketamine is frequently utilized due to the ability of SVR to be maintained at the expense of only a small increase in PVR. Etomidate combined with an opioid can also preserve the SVR-PVR balance during induction of anesthesia. The only interventions that are capable of lowering PHT without accompanying systemic vasodilation are: hyperventilation, and inhaled nitric oxide or prostanoids. With ventilation, the goal is to lower the $PaCO_2$ whilst utilising the lowest possible mean intrathoracic pressure. It is important to remember that the duration of inspiration has more influence on mean intrathoracic pressure than peak inspiratory pressure.

Persistent Left Superior Vena Cava (LSVC)

A LSVC is occurs in approximately 0.5% of the general population and 10% of CHD patients. In the majority of cases the LSCV drains into the coronary sinus. The use of cardiopulmonary bypass necessitates appropriate drainage of the coronary sinus and LSVC. Not surprisingly, this lesion renders retrograde cardioplegia non-functional. Left-sided central venous catheters and other transvenous procedures carry a risk of coronary sinus injury or thrombosis.

Hypertrophic Cardiomyopathy (HCM)

Hypertrophic cardiomyopathy is a rare autosomal dominant condition with variable penetrance and expressivity that affects 0.05 to 0.2 % of the population. It is characterized by inappropriate and usually asymmetrical LV hypertrophy. There is an initial peak incidence in the second decade of life and a later incidence that can span the third to sixth decades. Although LV outflow tract obstruction (LVOTO) resulting from hypertrophy in the interventricular septum (previously referred to as idiopathic subaortic stenosis – IHSS or asymmetrical septal hypertrophy) is the most common and well known form, it is now appreciated that the hypertrophy can occur anywhere in the LV. The obstructive forms produce midsystolic LVOTO related to the hypertrophy-induced narrowing of the LVOT combined with Venturi-induced systolic anterior motion of the mitral valve. Whilst most patients are asymptomatic there is a high incidence of ventricular dysrhythmias leading to sudden death that is often associated with sports or other vigorous activities (especially preadolescents and adolescents). Atrial dysrhythmias may occur in addition to the malignant ventricular dysrhythmias. Near-misses may present as syncope or presyncope. Although systolic function is frequently well-preserved, diastolic dysfunction coupled with mitral regurgitation often leads to heart failure. The dynamic LVOTO is worsened by sympathetic stimulation, hypovolemia and systemic vasodilation. Myectomy carries a risk of iatrogenic VSD, or injury to the aortic or mitral valves. There is also a propensity to recurrence as well as a risk of incomplete resection.

Transplantation

Cardiac transplantation in adults with CHD is has been utilized in the management of adults with end-stage CHD associated with severe systemic (morphologically right) ventricular dysfunction in l-TGA or in d-TGA after Mustard or Senning procedure, severe Ebstein's anomaly, failing Fontan circulation, and Eisenmenger syndrome (may require combined heart-lung transplant).

Additional Comments Pertaining to Anesthetic Care of ACHD

Preoperative Assessment

Obtaining an accurate history can be problematic as only one-half to three-quarters of adults with CHD can correctly name or describe their diagnosis. Further, by limiting their exercise, ACHD patients may be relatively asymptomatic or have grown accustomed to their condition. Assessment should look for evidence of long-term cardiac complications such as: pulmonary hypertension, ventricular dysfunction, residual shunts, regurgitant or stenotic valves, hypertension, aneurysms, conduction defects and dysrhythmias. As poor general health is also a risk factor, it is important to evaluate chronic non-cardiac co-existing diseases and/or complications of

ACHD such as: erythrocytosis, developmental and central nervous system disorders (seizures, previous ischemic/embolic events), chronic lung disease, cholelithiasis, and nephrolithiasis.

Investigation and Prognostication

In addition to assessing the anatomy, investigations are helpful in evaluation of the patient's functional class and the assessment of ventricular performance. Patients may undergo exercise testing, Holter monitoring, electrophysiologic studies, transthoracic and/or transesophageal echocardiography (TEE), and cardiac MRI. Cardiac catheterization is helpful: 1) in the investigation of pulmonary vascular disease and the determination of its responsiveness to oxygen and vasodilators, 2) in the assessment of functional suitability for Fontan completion, 3) as a supplement to echocardiography and MRI in the delineation of complex congenital lesions, and 4) in the assessment of the coronary arteries. Often this information is reviewed by a multi-disciplinary team in order to determine the prognosis and determine the timing, risks and benefits of planned surgical or cardiac catheterization interventional procedures. Major independent cardiac risk factors include: 1) pulmonary hypertension (three-fold risk increment), 2) cyanosis (higher mortality and more postoperative complications), 3) poor ventricular function, 4) dysrhythmias and 5) reoperative surgery. Risk is also increased by the presence of co-existing diseases such as hypertension, diabetes, and coronary artery disease.

Electrophysiological Considerations

In ACHD the congenital anomalies themselves as well as changes attributable to chamber dilation, fibrosis, surgical incisions, and hemodynamic deterioration contribute to the development of atrial and ventricular dysrhythmias. The occurrence of these dysrhythmias is a poor prognostic sign. Sinus node dysfunction, poor ventricular function and the desire for pregnancy may complicate pharmacologic management. Thus ACHD patients often present for procedures such as catheter ablation, open ablation (Cox-Maze procedure), as well as the insertion of automated implantable cardioverter defibrillators (AICDs) and anti-tachycardia pacemakers. In ACHD, epicardial lead placement may be required for AICDs and pacemakers as transvenous placement may be complicated by difficult venous access, unacceptable thrombosis risk, or the presence of intracardiac shunts.

Reoperation

With reoperation, re-opening the sternum can be dangerous due to the possibility of adhesions between the heart, great vessels (or conduit) and the sternum and the lack of a retrosternal space. Further, the adhesions obscure anatomic landmarks including the route of the coronary arteries. The presence of collateral vessels may also complicate dissection and increase the risk of hemorrhage. Patients who have undergone previous cardiac surgeries may also have sustained long-term myocardial damage.

General Principles of Anesthetic Management

Creation of the optimal anesthetic plan relies upon the anethesiologist's ability to correctly understand the patient's pathophysiology. Due to the variety of CHD lesions and the variations within a given type of CHD there is no "one-size-fits-all" selection of anesthetic medications. Rather the choices must be tailored to unique features of each case. Premedication should be selected carefully and its use particularly with opioids may be risky in patients in who further hypoventilation and hypoxemia could lead to decompensation. In patients with erythrocytosis, it is advisable to maintain preoperative hydration either by ensuring a minimum preoperative clear fluid fasting interval or by administering preoperative intravenous (IV) fluids. Intravenous fluid administration requires vigilance to avoid systemic embolization of air bubbles. Previous cardiac and non-cardiac procedures and hospital admissions may limit the availability of central and peripheral vascular access sites.

Monitoring

Previous surgery may also limit monitoring access sites. A Fontan repair precludes central venous access to the systemic atrium and ventricle. A BT shunt may render the invasive and non-invasive BP diminished or unobtainable in the corresponding upper limb. Selection of invasive monitoring requires a risk-benefit analysis that takes into account the patient's cardiac defect and overall health status as well as the planned surgical or cardiac catheterization procedures. Pulmonary arterial catheters are rarely used as 1) placement may be difficult or impossible, 2) there may be an excessive risk of dysrhythmias, and 3) the measurements may not be useful in guiding therapy. With open heart surgery, it is much more common to place intrathoracic catheters into otherwise inaccessible atria (LA or systemic atrial lines). Not only does TEE have utility in preoperative assessment (anatomy, ventricular function and remodelling, volume status), but its routine intraoperative use can provide a continuous assessment of ventricular function, preload and changes in valve function. Transesophageal echocardiography has also shown utility by uncovering and assessing unexpected pre-surgical findings in the OR in addition to its ability to evaluate the adequacy of the surgical repair. In CHD pulse oximetry is useful for tracking changes in pulmonary blood flow and R→L shunting in cyanotic patients. End-tidal CO_2 underestimates $PaCO_2$ in patients with R→L shunts.

Antibiotic Endocarditis Prophylaxis

In contrast to the more wide spread use of antibiotic prophylaxis in the past, current recommendations limit use to patients with: previous endocarditis, unrepaired cyanotic CHD including those with palliative shunts and conduits, the presence of prosthetic material or device during the first 6 months after the procedure, and residual defects at the site or adjacent to the site of a prosthetic patch or device. With the exception of these listed conditions, antibiotic endocarditis prophylaxis is no longer recommended for other forms of CHD. In cardiac surgery anti-staphylococcal (i.e. cefazolin) surgical wound antibiotic prophylaxis remains standard practice.

Anesthesia for Cyanotic Patients

Some features of managing cyanotic lesions have been mentioned previously whilst discussing specific lesions. Anesthesia management is aimed at promoting pulmonary blood flow through maintaining a favourable PVR, ensuring optimal driving pressure for pulmonary perfusion (systemic arterial or venous pressure depending upon the physiology), decreasing R→L shunting through maintaining normovolemia and SVR, sustaining a higher hematocrit, and modulating dynamic RVOTO, if present, with β-blockers.

The following additional general points are worth considering. Altering FiO_2 will have little effect on SpO_2 whilst, systemic vasoconstriction (phenylephrine, norepinephrine) will increase SpO_2 either by decreasing R→L shunting or by improving the flow through an aortopulmonary shunt or aorto-pulmonary collaterals. As well, cyanotic patients have a blunted hypoxic ventilatory response whilst the response to hypercapnia is preserved. Inhalational induction and subsequent titration (important in the case of relative excessive administration) is slowed in cyanotic patients. Intravenous medications theorectically have a more rapid, but perhaps not clinically significant onset. As mentioned previously, end-tidal CO_2 monitoring will underestimate $PaCO_2$.

Chronic hyperventilation and increased hematocrit are adaptations to improve oxygen transport in cyanosis. Erythropoietin overproduction leads to increased mass and stiffness of the red blood cells (RBC) which in turn results in elevated viscosity that increases cardiac work in hearts that already have chronic hypoxia-induced systolic and diastolic dysfunction. Erythrocytosis in the presence of iron deficiency results in poorly deformable RBCs that raise the risk of thrombosis.

Cyanotic patients have a variety of other coagulation abnormalities including abnormal platelet function, diminished platelet survival due to peripheral consumption, reduced levels of vitamin K-dependent coagulation factors as well as factor V and von Willebrand factor. Not only is erythrocytosis associated with an elevated INR and PTT, but this condition may interfere with the standard laboratory tests used to measure these indices. It is speculated that hyperviscosity and sluggish blood flow

are the reasons that the bleeding time remains normal despite these changes. In erythrocytosis there may also be a bleeding diathesis that results from tissue vascularity and dilated arterioles due to the release of endothelium-derived nitric oxide and prostaglandins as a result of the elevated wall sheer stress. The burden of the additional heme ring turn-over contributes to an increased incidence of cholelithiasis.

Chronic cyanosis is associated with end-organ dysfunction. In the heart, diastolic compliance is reduced and myocardial reserve is diminished. In the kidney, proliferative glomerular lesions and basement membrane thickening result in proteinuria and elevated creatinine and uric acid. Cyanotic patients also have an increased incidence of intracerebral abscesses.

Pulmonary Hypertension

In pulmonary hypertension (PHT) pulmonary vascular resistance increases as a result of changes in the pulmonary vascular bed that are the results of being subjected to increased pulmonary blood flow and near systemic blood pressure. The vascular bed undergoes changes that initially are reactive and reversible, but eventually become fixed and permanent. Pathological examination reveals media hypertrophy in small muscular arteries and arterioles, cellular proliferation in the intima, migration of smooth muscle cells into the subendothelium, and ultimately progressive fibrosis leading to obliteration of these vessels. Pulmonary hypertension is defined as a mean resting PAP > 25 mmHg or an exercise PAP > 30 mmHg and as a PVR > 300 dynes/s/cm^5. The reactive nature of PHT is characterized by exaggerated responses to sympathetic stimuli (i.e., pain or surgical stress response), acidosis, hypercarbia, hypoxia, and hypothermia. Although it may be easier and preferable to prevent exacerbations of PHT than it is to treat them, the strategies used are similar. Management of ventilation includes the use of high FiO_2 and hyperventilation with the use of the minimal possible intrathoracic pressure aiming for a $PaCO_2$ of 25-30 mmHg and a pH above 7.45. One must aim for an anesthetic depth that will blunt the sympathetic response as best as possible. The choice of anesthetic medication used to achieve this goal will depend upon the particular circumstances, but opioids (sufentanil, fentanyl, remifentanil) are often helpful. Inotropic support with β-stimulators (dobutamine or isoproterenol) or phosphodiesterase-5 inhibitors (milrinone) may permit the patient to tolerate this depth of anesthesia and help the RV to cope. However, one must beware of the systemic hypotension that may accompany their use and aggravate R→L shunting. In fact systemic vasoconstriction with norepinephrine or phenylephrine may be essential in order to increase coronary perfusion pressure and relieve sub-endocardial ischemia and improve ventricular function. Selective pulmonary vasodilation with inhaled nitric oxide is a mainstay of current therapy although aerosolized prostanoids have also been used with success. Oral sildenafil has been used for both prophylactic and maintenance therapy in PHT. Other medications with vasodilating effects such as magnesium sulphate, antiendothelins, antiangiotensins and calcium channel blockers has been used in PHT, with their utility limited by their systemic vasodilating properties.

Poor Ventricular Performance

Impaired ventricular function is common in ACHD. In the absence of surgical correction, ventricular remodelling such as concentric hypertrophy and spherical dilation worsens over time. In general pressure overload is less well tolerated than volume overload. Cyanotic patients are at additional risk of ventricular dysfunction and myocardial ischemia as consequences of impaired oxygen transport, decreased diastolic perfusion pressure caused by shunts or collaterals, and from impaired or occluded myocardial microvascular perfusion as a result of erythrocytosis and hyperviscosity. Coronary artery disease may be an additional detrimental factor in ACHD.

Pregnancy

With the risks imposed by pregnancy for forms of ACHD, contraceptive counselling is essential. Management of pregnancy in patients with moderate to severe ACHD requires a multidisciplinary team. The normal physiological changes of pregnancy (50% increase in blood volume 50%, 30-40% increase in cardiac output, 30% increase in heart rate, and decreased SVR) may lead to pulmonary overload in patients with L→R shunts and to increased shunting and cyanosis in R→L shunts. Maternal adaptation to the postpartum physiological changes also carries a particularly high mortality risk. Risk factors for elevated maternal morbidity and mortality include: cyanosis, pulmonary hypertension, poor functional status and/or heart failure, poor dysrhythmias control, LV obstructive lesions, and prior cerebral ischemic events. Not only does cyanosis carry approximately a 50% maternal morbidity and mortality rate, it is also a major risk factor for fetal and neonatal complications. Maternal mortality in the presence of PHT (i.e. Eisenmenger's syndrome) is estimated at between 30 and 70 percent. The normal physiological adaptation to pregnancy is altered with coarctation of the aorta with an increased heart rate being necessary to support most of the increase in cardiac output. Complications (ventricular failure, dysrhythmias, endocarditis, aortic dissection and thromboses) may arise in almost one third of pregnant patients with coarctation of the aorta.

In pregnant ACHD patients with L→R shunts, the lower SVR with spinal or epidural analgesia or anesthesia may be beneficial. In contrast, in pregnant patients with palliative L→R shunts general anesthesia is often recommended as it is essential to avoid dropping the SVR. For reasons mentioned above, it is necessary to maintain the tachycardia in pregnant patients with coarctation of the aorta. Intrathecal anesthesia is contraindicated in the presences of aortic coarctation. In the parturient with unrepaired TOF a general anesthetic may be easier to manage, but epidural anesthesia has been used with gradual onset of the sympathetic blockade. Phenylephrine and esmolol must be available for rapid treatment of a TOF spell). The rapid onset of sympathetic block contraindicates spinal anesthesia in the parturient with Eisenmenger syndrome. A cautiously conducted general anesthesia with normocarbic ventilation is preferred, although with careful maintenance of preload and afterload, a gradually induced epidural anesthetic may be possible in this extremely high risk group.

Suggested Reading:

Cannesson M, Earing MG, et al. Anesthesia for noncardiac surgery in adults with congenital heart disease. Anesthesiology 2009; 111:432-40
Chassot PG, Bettex DA. ,Anesthesia and adult congenital heart disease. J Cardiothoracic Vascular Anesthesia, 2006; 20:414-437
Lovell AT. Anaesthetic implications of grown-up congenital heart disease. Br J Anaesth 2004; 93:129-39
Summary of recommendations: Care of the adult with congenital heart disease. J Am Coll 2001; Cardiol 37:1161-1198