Anesthesia for miscellaneous cardiac lesions

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Introduction

This chapter will discuss the anatomy, pathophysiology, surgical approach, and anesthetic management of two groups of rare lesions: vascular rings and anomalies of the coronary arteries. Mitral regurgitation (MR) and anesthetic considerations for pericardial effusion and tamponade will then be reviewed.

Vascular rings

Vascular rings are a variety of anomalies of the aortic arch and its branches, which result in compression of the trachea and/or esophagus. These lesions are rare, accounting for less than 1% of all congenital heart defects.

Anatomy

Vascular rings were first described in 1737 by Hommel who described a double aortic arch. Bayford reported the first case of retroesophageal right subclavian artery in 1794. However, it was not until 1945 that a vascular ring was successfully divided by the pioneering efforts of Robert Gross.

Vascular rings encompass many different vascular anomalies, all of which result from the abnormal regression of the aortic arch complex. The majority (60%) of all vascular rings are of the double aortic arch variety, which results from the persistence of the fourth aortic arch. Many variations in the arrangement of the aorta and its branches exist, and can result in complete or partial rings.

Of the many different anatomies, (i) double aortic arch and (ii) right aortic arch with aberrant left subclavian artery are the most common. In double aortic arch, the right aortic arch passes to the right of the esophagus to join the left-sided descending aorta, thus completing the vascular ring (Fig. 23.1). Two other varieties of vascular ring include (iii) right aortic arch with mirror image branching and left ligamentum arteriosus, and (iv) left aortic arch with retroesophageal right subclavian artery.

Intracardiac lesions are rarely seen with double aortic arch but are often present in cases of (v) left aortic arch with right descending aorta. The left arch crosses behind the esophagus, and a right ligamentum arteriosus completes the ring.

Partial vascular rings or “slings” may also occur. For example, (i) an aberrant right subclavian artery of an otherwise normal arch may pass to the right behind the esophagus with resultant dysphagia. Other partial rings that have been described include (ii) ductus arteriosus sling and (iii) compression of the lower trachea from severe malrotation of the heart. In the latter two, dividing the ductus arteriosus relieves the compression.
Finally, with the pulmonary artery sling, the left pulmonary artery arises from the proximal right pulmonary artery, and passes behind the trachea. The ligamentum arteriosum completes the vascular ring by compressing the trachea anteriorly.

Pathophysiology and natural history

Symptoms are usually prominent in patients with a tightly obstructed ring as is seen in double aortic arch. Infants often present with respiratory distress, stridor, and swallowing difficulties within the first 6 months of life. Subcostal retractions can be seen in severe obstruction. Recurrent respiratory difficulty or dysphagia in a young infant should raise the question of the presence of a vascular ring. However, the diagnosis of vascular ring in infants without associated anomalies is often delayed months to years after onset of symptoms. Partial vascular rings may be asymptomatic if there is little tracheoesophageal compression. Older infants and adults with undiagnosed vascular rings have presented with acute esophageal foreign body impaction, unsuccessful treatment of the esophagus or trachea. Computed tomography (CT) and magnetic resonance imaging (MRI) have been shown to be diagnostically accurate, eliminating the need for exploratory surgery, though unnecessary and rarely done, may reveal a pulsating mass compressing the esophagus or trachea. Computed tomography (CT) and magnetic resonance imaging (MRI) have been shown to be diagnostically accurate, eliminating the need for angiography. Echocardiography is used liberally by some centers to evaluate the presence of a congenital heart anomaly, which may be present in up to 20% of children with symptomatic vascular rings.

Surgical approaches

Delayed treatment may result in tracheobronchial damage in symptomatic patients. Surgery, however, is not indicated if symptoms are mild or absent. Best exposure is provided by the left thoracotomy approach through the fourth intercostal space. If coexisting cardiac anomalies require repair, a median sternotomy may be used. The arch, including the retroesophageal component is dissected out completely. Care must be given to the identification and division of the ring without compressing blood flow to the descending aorta or carotid arteries. In double aortic arch, the non-dominant arch is divided and sutured at its distal end close to the junction with the descending aorta. The ligamentum arteriosus is also divided in all cases. The trachea and esophagus are dissected and freed of all strands or bands of tissue that may add to the constriction. On occasion, the descending aorta is suspended to the rib periosteum to keep it away from the esophagus. If the vascular ring is of the right aortic arch type, the division of the left ligamentum arteriosus opens the ring and relieves the constriction.

On rare occasion, right thoracotomy may be indicated. This is the case in double aortic arch with a smaller non-dominant right aortic arch, as well as the case of left aortic arch with a retroesophageal subclavian artery and right ligamentum arteriosus.

Video-assisted thoracoscopic division of vascular rings has been described. This approach is limited by decreased vascular exposure and the reduced ability for direct vascular control. The safety of the thoracoscopic approach is increased if there is an absence of blood flow and atresia of the ring structure undergoing division.

The pulmonary artery sling is usually repaired via median sternotomy with cardiopulmonary bypass (CPB). The left pulmonary artery is removed from its origin on the right pulmonary artery, brought in front of the trachea, and reimplanted on the main pulmonary artery.

Anesthetic considerations and approach

With any surgery involving dissection and ligation of large vascular structures, there is a potential for significant and rapid blood loss, necessitating adequate and reliable intravenous access. Induction of anesthesia is straightforward in the less symptomatic patient. However, children with significant airway compression are at risk for complete airway obstruction and benefit from an inhalation induction with the maintenance of spontaneous ventilation. In these symptomatic patients, paralysis should be administered only after the ability to assist with positive pressure ventilation has been ascertained. These patients may also require a smaller than expected endotracheal tube size. Neuraxial opioids and/or local anesthetics, either by single shot caudal, or continuous techniques, may greatly facilitate pain relief, early extubation, and pulmonary toilet. In addition to standard monitors, an arterial catheter should be placed in most of these patients because of the potential for hemodynamic and respiratory instability. In the case of an aberrant subclavian artery, the site of the arterial catheter should be chosen after discussion with the surgeon of the surgical approach. Central venous catheterization should be considered for extensive surgery, poor vascular access, or anticipated hemodynamic instability. Consideration should also be given to monitoring the cerebral circulation with near infrared spectroscopy and/or transcranial Doppler ultrasound in cases where cerebral blood flow may be compromised, e.g. clamp-
ing and reimplantation of a carotid artery that arises from an aberrant subclavian artery. Finally, some of these operations may be facilitated by single lung ventilation to improve surgical exposure and lessen movement of vascular structures with ventilation. See Chapter 16 for a discussion of the available techniques.

**Postoperative airway management and pain control**

Asymptomatic patients can be extubated at the end of the case. Respiratory symptoms may worsen in symptomatic patients during the first postoperative week, occasionally necessitating intubation for adequate pulmonary toilet. For these infants, continuous positive airway pressure with humidified gas via nasal prongs may also be helpful. Good postoperative analgesia is essential to encourage deep breathing and lung expansion. This can be facilitated with epidural analgesia, intercostal rib blocks or adequate intravenous opioid administration.

**Anomalies of the coronary arteries**

Abnormalities can exist in the number, origin, and termination of the coronary arteries. The number of coronary arteries can vary from one to four, often occurring in association with other congenital defects. A single coronary artery may be associated with myocardial ischemia, myocardial infarction or sudden death. Coronary arteries may have an anomalous origin from the aorta, the innominate artery, the carotid artery, the left anterior descending artery or, most commonly, from the pulmonary arteries.

**Anatomy**

**Anomalous origin from the aorta**

If the left main coronary artery arises from the right aortic sinus, it courses between the ascending aorta and the pulmonary artery where compression can occur, leading to myocardial infarction or sudden death. Variations in the aortic origin of the coronary arteries often occur in association with congenital heart defects. In 7% of patients with tetralogy of Fallot, the left anterior descending artery originates from the right coronary artery. This repair is straightforward because of the anterior origin of the anomalous artery and its close proximity to the aorta.

**Pathophysiology and natural history**

The physiologic changes produced by ALCAPA worsen after delivery. In utero, when the pulmonary artery pressure and oxygen saturation nearly equal the systemic pressure and oxygen saturation, left ventricular myocardial perfusion and oxygenation are adequate. Myocardial ischemia develops soon after birth as pulmonary vascular resistance (PVR) falls, causing a marked decrease in left coronary artery perfusion.
pressure. The infant’s survival is dependent on the extent of collateral formation from the right coronary artery to the left coronary artery. These intercoronary collaterals, however, also allow the flow of blood from the right coronary artery via the left coronary artery system into the pulmonary artery and are often referred to as coronary artery fistulization. This coronary artery “steal” causes lower perfusion pressure and results in myocardial damage.

Typical presentation includes profuse sweating, tachycardia, tachypnea, dyspnea, coughing, wheezing, pallor, and failure-to-thrive. In some infants atypical chest pain upon eating and crying has been mistaken for colic.23 One should have a high index of suspicion of ALCAPA in any infant with global myocardial dysfunction. However, about 10% of patients with ALCAPA with good collateral flow do not develop myocardial ischemia until adolescence or adulthood.24 Adults have presented with malignant ventricular arrhythmias,25 shortness of breath with exercise,24 cardiac murmur,26 and cardiac arrest during exercise.27 Although the most common cause of sudden death in young competitive athletes is hypertrophic cardiomyopathy; 13% of deaths in these athletes involve anomalous coronary artery origin.28 All older patients with asymptomatic ALCAPA had multiple unusual color flow Doppler signals within the ventricular septum, representing septal coronary collaterals.29

Physical examination reveals evidence of congestive heart failure, cardiomegaly and the murmur of mitral insufficiency. The chest radiograph consistently shows massive cardiomegaly. Bronchial compression by the enlarged heart can result in atelectatic changes in the left lung.

The electrocardiogram (ECG) is abnormal in all patients, showing evidence of ischemia, infarction and left ventricular hypertrophy. Electrocardiogram findings have been described that are present in all patients with ALCAPA but absent from most patients with myocarditis and cardiomyopathy. These ECG criteria are: (i) Q-wave depth greater than 3 mm; (ii) Q-wave width greater than 30 ms; and (iii) a QR pattern in one of the following leads: I, aVL, V5–V7.30

Echocardiography can demonstrate the anatomic origin of the ALCAPA and provides an assessment of the degree of left ventricular impairment. Studies show a significant enlargement of the right coronary artery and a dilated left ventricle with global hypokinesia. Pulse and color flow Doppler imaging can directly visualize the anomalous origin as well as the reversal of flow from the ALCAPA into the pulmonary artery.19 Cardiac catheterization is not routinely performed unless ALCAPA is suspected but cannot be visualized by echocardiography. When performed, aortography can show filling of the left coronary artery through collaterals from the dilated right coronary artery and can exclude other anomalies.

The need for early surgical repair of all infants with ALCAPA is essential in even asymptomatic infants because of the extremely poor survival with medical management. Ninety percent of undiagnosed or medically treated infants die within the first year of life. Sudden death frequently occurs in untreated older children and adults. Thus, surgical correction is indicated in all patients with ALCAPA.

Surgical approaches

Surgical treatment for ALCAPA is directed towards correcting the “coronary steal” phenomenon, and increasing left ventricular myocardial perfusion and function. This can be accomplished by either reconstituting a two-coronary system or by simply ligating the fistulous flow. Restoring a two-coronary circulation is preferred, and when possible, direct coronary–aortic reimplantation is performed.31 Scarred myocardium or free wall aneurysm is not addressed at the time of initial surgery.

Simple ligation of the ALCAPA eliminates the “steal” phenomenon, and in the past had been recommended as the procedure of choice in critically ill infants but has a prohibitive mortality rate ranging from 20% to 50%.32 A single coronary artery system is less physiologic with greater risk of postoperative complications, a higher early postoperative mortality, and a higher potential for atherosclerosis as well as late sudden death.

Surgical reconstitution of a two-coronary artery system results in greater recovery of left ventricular function and is now the standard surgical procedure. Direct coronary aortic reimplantation is an excellent procedure because it is simple, does not require prosthetic material, and is expected to provide excellent late results. However, it often requires creative surgical technique to obtain sufficient length and correct angling of the coronary artery to the aorta (Fig. 23.2). This is difficult when the anomalous vessel originates from the left posterior wall of the pulmonary trunk. If mobilization and reimplantation will compromise the vessel, the left subclavian artery may be anastomosed to the left coronary artery (Fig. 23.3). This procedure may be done without CPB but is not feasible if the left main coronary artery is short in length.33

A saphenous vein bypass graft may also be used, but requires deep hypothermia (18°C) with total circulatory arrest. The small vein caliber and the incidence of late graft occlusion limit the utility of the saphenous vein graft in infants. Alternatively, an aortocoronary bypass graft with a free segment of the subclavian artery may be used. An end-to-side retroaortic coronary bypass graft construction with a free segment of the left subclavian artery is applicable in the majority of infants. More recently, the left internal mammary artery (LIMA) has been used but long-term results are lacking with ALCAPA.34,35

Newer techniques utilize the pulmonary artery as a conduit graft from the left coronary artery to the aorta. This is useful for cases where the left coronary artery arises from the left posterior sinus.36 The Takeuchi procedure37 uses a flap derived from the anterior wall of the pulmonary trunk to
be reported with the Takeuchi procedure. Tashiro et al.\textsuperscript{38} has described left coronary angioplasty using pulmonary trunk without prosthetic material, which is expected to yield excellent late results. This procedure combines left coronary

create a coronary tunnel inside the pulmonary trunk between a surgically created aortopulmonary window and the left coronary ostium (Fig. 23.4). The opening in the pulmonary trunk is then patched with pericardium. Late complications have
angioplasty, side-to-side anastomosis of the aorta with the newly created left coronary artery and direct anastomosis of the transected pulmonary artery. To date, no difference has been shown in the long-term left ventricular function or late mortality among the various surgical techniques which re-establish a two-coronary circulation.39

There have been conflicting opinions as to the necessity for the repair of mitral incompetence in these patients. Some surgeons recommend mitral annuloplasty at the time of the initial operation.40 However, the vast majority of regurgitant valves gradually improve within 6 months by serial echocardiography without annuloplasty. Even severe MR has been reported to regress fully after reperfusion alone in 62% of cases.41 Many surgeons now feel that mitral valve repair is not generally necessary at the time of the initial operation.42 In some patients, the persistence or recurrence of MR may signify a significant coexistent coronary stenosis. Patients who have significant obstruction in their left coronary artery will have residual MR and may require not only mitral valve repair, but also revascularization of the left coronary artery.42

Anesthetic considerations and approach

Infants with ALCAPA are often critically ill with little cardiac reserve and significant ischemia. More severe preoperative MR is associated with increased perioperative mortality.43 Adequate monitoring, including a multilead ECG, arterial pressure monitoring, and central venous access for drug administration and assessment of volume status are essential. Induction should be a gradual one to avoid major swings in blood pressure. A gentle and rapid laryngoscopy is also critical. Fluid administration is titrated to assure adequate preload for maintenance of cardiac output (CO) while avoiding pulmonary edema. Measures to mildly increase PVR such as normocapnia and decreasing FiO2 to the lowest level tolerated can help minimize the coronary steal phenomenon. Inotropic agents can improve cardiac function, but can also increase heart rate (HR) and myocardial oxygen consumption and worsen the ischemia. The cardiovascular depressant effects of volatile anesthetics are often poorly tolerated, and an opioid technique may be preferred.

If repair requires the use of CPB, significant post-bypass inotropic support may be needed, e.g. dopamine, dobutamine, or epinephrine. Nitroglycerin is often used to improve coronary perfusion. Decreasing the afterload of the left ventricle is also desirable and can be accomplished with an inodilator (milrinone) or a vasodilator (sodium nitroprusside). Mechanical support of the left ventricle with a left ventricular assist device (LVAD) may be required in some patients who are unable to be weaned from CPB.44 This may be more common in the younger infant with poor collateralization of coronary blood flow and acute myocardial infarction. Postoperative pain control with intravenous narcotic is adequate. Most patients are kept intubated and ventilated postoperatively to allow time for ventricular recovery.

Normalization of LV function occurs substantially after restoration of a two-coronary circulation, although this may take as long as 2 years, and some degree of chronic impairment may persist. Even in the group requiring LVAD support, a high survival rate and good long-term recovery can be achieved.

The optimal follow-up method after ALCAPA repair is controversial, with ECG, Holter monitoring, stress-thallium scanning, and cardiac catheterization all showing equivalent results. Serial echocardiography can be useful to assess the left ventricular function, the severity of mitral insufficiency, and the patency of the revascularized left coronary artery flow. Long-term survivors of ALCAPA repair show regional impairment of myocardial flow reserve which may contribute to impaired exercise performance.45 However, even under stress testing, the normal growth of the heart has not been found to compromise the anastomosis. Patients with ALCAPA that survive the perioperative period have an excellent prognosis for functional recovery of the left ventricle regardless of their preoperative state.46

Mitral regurgitation

Mitral regurgitation rarely occurs as an isolated lesion in congenital heart disease. Rather, it usually occurs as a component of another lesion, such as complete or partial atrioventricular canal defect, mitral valve prolapse, myocardial or papillary muscle infarction such as that seen with the ALCAPA syndrome (see above), connective tissue disorder such as Marfan’s syndrome, rheumatic disease, endocarditis, or Kawasaki disease.47,48

Pathophysiology and natural history

In the normal mitral valve, the leaflets are asymmetric, with the anterior leaflet spanning one-third of the annulus, and the C-shaped posterior leaflet two-thirds. The anterior leaflet inserts into the anterolateral papillary muscle, and the posterior leaflet into the posteromedial papillary muscle. The papillary muscles insert into the left ventricular free wall in the normal mitral valve; the anterior leaflet is normally perfused by the left coronary artery and the posterior leaflet by branches of both left and right coronary arteries.49 Valve competency depends on the large posterior leaflet overlapping the anterior leaflet during systole, and thus complete coaptation is necessary. Any process interfering with this coaptation, be it a cleft in the posterior leaflet, annular dilation from left ventricular infarction or severe dysfunction, may cause MR.

The clinical severity and symptoms of MR can be classified as mild, moderate, or severe.50 In the patient with chronic MR, the patient may be asymptomatic. As MR worsens, both
the left atrium and left ventricle dilate, resulting in enlargement of the mitral annulus, and creating a vicious cycle of worsening MR as the annulus dilates. Resting HR increases, and as left atrial pressures (LAPs) increase, pulmonary venous, capillary, and finally pulmonary artery hypertension can occur. This results in tachypnea, pulmonary edema, poor feeding and diaphoresis in infants, and may result in atrial arrhythmias such as atrial fibrillation. The enlarged left atrium may cause compression of the left mainstem bronchus in infants. Obstructive airways disease and frequent infections occur in infants with moderate to severe MR. Physical examination in moderate to severe MR often reveals a diaphoretic, tachypneic patient, with resting tachycardia and an increased precordial impulse. The left ventricular apex may be displaced laterally, and the second heart sound intensity may be increased with pulmonary hypertension. A holosystolic, high frequency murmur is heard at the apex, radiating to the axilla. A low pitched diastolic rumble is heard with moderate or severe MR. The presence of a third heart sound indicates severe MR. Chest radiograph reveals cardiomegaly, with left atrial and left ventricular enlargement, and varying degrees of increased pulmonary vascular markings. The left lower lobe may have atelectasis due to left atrial enlargement and compression of left sided bronchi. Medical treatment includes the use of diuretics, digoxin, and afterload reduction is provided by angiotensin-converting enzyme inhibitors. Acute MR, such as that seen with acute ALCAPA, is often very poorly tolerated, with rapid development of pulmonary edema and respiratory and circulatory decompensation. Indications for surgical intervention include uncontrolled congestive heart failure resulting in failure to thrive, progressive enlargement of the left atrium and ventricle despite medical management, or atrial arrhythmias or persistent airway symptoms.

Echocardiographic diagnosis is essential to define the anatomy underlying the MR, and its severity. Various schema for grading echocardiographic severity have been devised; generally, when there is flow reversal in the pulmonary veins, the MR is considered significant. A regurgitant fraction can be calculated. Another indicator of severity of MR is the diameter of the regurgitant jet at the level of the orifice. Cardiac catheterization is infrequently required for MR, but a regurgitant fraction can often be calculated angiographically, with a 20% regurgitant fraction considered mild MR, 20–40% moderate, 40–60% moderately severe, and greater than 60% severe. The LAP is elevated with a large A wave.

Surgical approaches include annuloplasty with a prosthetic DeVega or Carpentier ring, suturing the cleft in the mitral valve, resection of a portion of the posterior leaflet, repairing or foreshortening a ruptured or damaged chordae tendineae, or mitral valve replacement. Mitral valve repair is preferred in most centers in growing children, because of the need for anticoagulation with prosthetic valves, and the need for repeated replacement until growth is complete. The surgery is done via midline sternotomy with CPB, and is often approached through an incision in the left atrium. The exact surgical approach is often not determined until the surgeon inspects the anatomy. After repair of the valve, appropriate annular size may be tested by passing a Hegar dilator of appropriate size for the patient's body surface area, and competence of the valve by injecting saline rapidly through the valve orifice into the left ventricle and noting competence while the heart is flaccid, which often predicts residual regurgitation.

**Anesthetic considerations**

The optimal hemodynamic state for a patient with moderate or severe MR consists of afterload reduction, adequate preload and contractility, and high–normal HR. Faster HRS lead to less diastolic filing time, and less time for ejection, which will lead to a smaller regurgitant fraction, i.e. more forward stroke volume and CO. Afterload reduction will encourage forward flow as well, and adequate preload is necessary for forward flow to be normal. Contractility should be maintained at normal levels to ensure ejection of the large stroke volumes seen in this lesion. A number of anesthetic regimens can be used to meet these goals, but high dose synthetic narcotics will need to be combined with a vagolytic agent such as pancuronium to maintain high HRS. Ketamine may not be desirable because this agent usually elevates systemic vascular resistance. Volatile agents are acceptable as long as they do not unduly depress contractility and they maintain HR; thus isoflurane, desflurane, or sevoflurane may be preferable to halothane in this lesion.

Monitoring consists of standard monitors, arterial and central catheters, and transesophageal echocardiography (TEE), which is critical to reconfirm the preoperative findings, and most importantly, to assess the adequacy of surgical repair. In addition, since the left side of the heart is opened for mitral valve surgery, TEE is critical to assess adequacy of intra-cardiac de-aging before the aortic cross-clamp is removed and before the patient is weaned from CPB. Carbon dioxide is insufflated into the surgical field in some centers when the left side of the heart is open to air to decrease the number and size of air bubbles in the heart. A left atrial catheter is often placed transthoracically by the surgeon during rewarming on bypass, in order to measure left sided filling pressures after bypass. Transcranial Doppler ultrasound is used in some centers to detect cerebral emboli, along with near infrared cerebral oximetry. Inotropic support is often required after CPB, and phosphodiesterase inhibitors such as milrinone are often used because of their vasodilating effects on both pulmonary and systemic circulations, and effects on both systolic and diastolic ventricular function.

Assessment of the postoperative repair and hemodynamics after bypass includes LAP measurement, both the absolute
number (may be elevated in both residual MR, or in mitral stenosis), and the presence or absence of a large V wave (present in MR, not as prominent in mitral stenosis or left ventricular dysfunction), which signifies residual MR. Once again TEE is crucial to determine the presence of residual MR, or the occasional creation of mitral stenosis, signified by elevated mitral valve inflow velocities or abnormal inflow patterns (see Chapter 9). These patients usually are not at great risk for postoperative bleeding because the suture lines are low pressure atrial sutures. Length of postoperative ventilation depends entirely on the patient’s pre and postoperative condition; some older patients with preserved ventricular function may be candidates for early extubation.

**Pericardial effusion and tamponade**

The pericardium is composed of visceral and parietal layers, and forms a fibroserous sac around the heart and extends a short distance onto the great vessels. Collagen and connective tissue fibers form the fibrosa, and are compliant under normal conditions of low fluid volume and stretch, but when fluid in the pericardial sac increases significantly, the steep portion of the pressure–volume curve may be reached, and intrapericardial pressures increase greatly, and create tamponade physiology. The pericardium is opened and partially removed for most cardiac surgeries, but the closed mediastinal space still has limited reserve to accumulate blood and fluid, especially in small infants.

Symptomatic pericardial effusion and tamponade may be seen in a number of clinical settings, both post-surgical and in medical conditions. Acute postoperative hemorrhagic tamponade is obviously a life-threatening emergency, but mediastinal bleeding and tamponade physiology may develop more slowly, over hours to several days postoperatively. Other causes of pericardial effusion and tamponade include cardiac perforation from cardiac catheterization or central venous catheter placement, chylos pericardial effusion after surgery, acute viral or bacterial infections, trauma, post-pericardiotomy syndrome, malignancy, congestive heart failure, renal failure, and inflammatory and autoimmune disorders.

Cardiac tamponade occurs when fluid, blood, or blood clots fill the pericardial space or mediastinum and increase pressure enough to significantly affect CO. Beck’s triad consists of hypotension, elevated systemic venous pressure, and a small quiet heart on auscultation. Clinically patients with tamponade physiology have dyspnea, tachycardia, distended neck veins, narrow pulse pressure, and pulsus paradoxus in the presence of a pericardial effusion. Tamponade physiology develops as right and left atrial, and biventricular end-diastolic pressures equalize as the cardiac chambers compete for restricted space. Diastolic filling and thus stroke volume become restricted, and the sympathetic nervous system compensates by increasing the contractile state, ejection fraction, and HR. Inspiration lowers intrathoracic pressure and promotes venous inflow into the right ventricle, which fills, but this shifts the interventricular septum to the left, restricting left ventricular filling. The negative intrathoracic pressure also decreases pressure in the pulmonary veins, and in combination with elevated left ventricular diastolic pressure, this also inhibits left ventricular filling, and thus stroke volume decreases greatly during inspiration, creating pulsus paradoxus. Interestingly, pulsus paradoxus detected on the pulse oximeter plethysmographic waveform correlates well with clinical cardiac tamponade in pediatric patients. Of note, positive pressure ventilation, atrial septal defect, severe left ventricular dysfunction, hypertrophic cardiomyopathy, and aortic insufficiency all reduce or eliminate pulsus paradoxus.

Aside from clinical signs and symptoms, echocardiography is the most important diagnostic tool in pericardial effusion and tamponade syndromes. The size and location of the effusion can be defined, as well as its consistency—serous or bloody vs. fibrinous or clots. In addition, tamponade physiology can be confirmed by detecting reduced mitral valve and pulmonary vein inflow during inspiration. The echocardiogram can also direct the physician to the best and safest location for pericardiocentesis, or open drainage.

In a patient with tamponade physiology, the induction of general anesthesia, muscle relaxation, tracheal intubation, and positive pressure ventilation are fraught with danger, and cardiac arrest and death may occur in this scenario. The patient is often barely compensating, and only the combination of maximal sympathetic stimulation and negative intrathoracic pressure during spontaneous respiration allow enough stroke volume to maintain barely adequate CO. Any upset in this balance can result in cardiac arrest. Anesthetic agents may remove sympathetic stimulation, and the institution of positive pressure ventilation may increase intrathoracic pressure to the point that systemic and pulmonary venous return essentially cease. The ideal situation would be to drain some of the fluid under local anesthesia so that ventricular filling and CO can improve to the point that the patient can tolerate anesthetic induction for a definitive procedure. This is often possible in the adult or cooperative older child or teenager, but not in the infant or toddler. In this situation, ketamine, despite its potential for direct myocardial depression, is usually well tolerated, while maintaining spontaneous ventilation until some fluid can be drained.

If general anesthesia must be induced, the cardiologist or surgeon responsible for the drainage of the fluid must be present, prepared to emergently access the pericardial or mediastinal space, either by needle or incision. Echocardiographic guidance is essential for pericardiocentesis, where the pericardial space is not being accessed under direct vision. All equipment and crossmatched blood must also be readily available, and in some instances it is prudent to have
the subxyphoid area steriley prepared and draped for immediate incision if hemodynamic collapse occurs on induction. An adequate period of preoxygenation is essential. Intravascular volume loading is recommended because it may maximize venous return, and is unlikely to worsen the situation acutely. Etomidate would appear to be the preferable agent for rapid intravenous induction of anesthesia because of its lack of negative inotropic effect on the myocardium. Ketamine would be a possible choice, but again has direct negative effects on the myocardium. If the patient is not at high risk for gastric aspiration, it may be preferable to induce anesthesia and keep the patient breathing spontaneously or gently assisted, if possible, until some of the fluid can be drained. Succinylcholine may need to be avoided because of its propensity to cause bradycardia. Tracheal intubation should be rapid, and positive pressure ventilation extremely gentle, or avoided for as long as possible. Preparations should be made for a full resuscitation, including epinephrine and atropine. In the event of hemodynamic collapse, drainage of the pericardial space must proceed immediately while resuscitative efforts are made. Draining the fluid normally allows the patient to recover enough CO to continue the procedure at a more controlled pace.

Drainage of the pericardium or mediastinum can be accomplished by pericardiocentesis, where the space is accessed under echocardiographic guidance with a needle, then a guidewire, and finally a catheter is placed for drainage. Injection of agitated saline as echocardiographic contrast medium may assist in localizing the pericardial space. This is normally performed by a cardiologist, and should be performed in a location where full resuscitative resources, including personnel, are available. A subxyphoid pericardial window for drainage is frequently performed by the surgeon, with a mediastinal drain left in place for several days. Occasionally, as in the case of constrictive pericarditis with impending tamponade, a pericardial stripping must be done with full sternotomy. Acute postoperative tamponade from mediastinal bleeding is heralded by low blood pressure and poor systemic perfusion accompanied by elevated central venous and LAPs. Mediastinal tube drainage may be increased, or may have decreased greatly, giving the team a false sense of security. A widened mediastinum may be seen on chest radiograph, and this can be confused with low CO due to myocardial dysfunction. Echocardiography, if time permits, will exclude the latter diagnosis. The sternum must be reopened immediately at the bedside in the intensive care unit in cases of impending cardiac arrest. A more controlled re-exploration may be undertaken in the operating room if time permits.

Reported complications for pericardiocentesis in pediatric patients range from death from cardiac perforation and tamponade to pneumopericardium, to ST changes from coronary artery lacerations. Higher complication rates are seen with younger patients under the age of 2 years, inexperienced operators, and lack of echocardiographic guidance, making the latter essential. Hand-held portable ultrasound technology has progressed to the point where this may be a viable option in any hospital setting.

Summary of management

Vascular rings

- Reliable vascular access is essential because of the potential for significant and rapid blood loss. Arterial line for most; also consider central line.
- An inhalation induction should be performed with maintenance of spontaneous ventilation until the ability to assist with positive pressure ventilation has been ascertained.
- Patients may require a smaller-than-expected endotracheal tube size, and single lung ventilation may be desirable.
- Minimally symptomatic patients can be extubated at the end of the case.
- Good postoperative analgesia is essential using epidural analgesia, intercostal rib blocks or adequate intravenous opioids.

Anomalies of the coronary arteries

- Infants with ALCAPA are often critically ill with little cardiac reserve and significant myocardial ischemia.
- Adequate monitoring, including a multilead ECG, arterial pressure monitoring, and central venous access for drug administration and volume assessment are essential.
- Induction should be gradual to avoid major swings in blood pressure. A gentle and rapid laryngoscopy is also critical.
- Fluid administration is titrated to assure adequate preload for maintenance of CO while avoiding pulmonary edema.
- Measures to mildly increase PVR such as normocapnia can help minimize the coronary steal phenomenon.
- Inotropic agents can improve cardiac function, but can also increase HR and myocardial oxygen consumption and worsen the ischemia.
- The cardiovascular depressant effects of volatile anesthetics are often poorly tolerated, and an opioid technique may be preferred.
- After CPB, significant inotropic, inodilator, and coronary and systemic vasodilator support may be needed.
- Most patients are kept intubated and ventilated postoperatively to allow time for ventricular recovery.
- Mechanical support of the left ventricle with a LVAD may be required in some patients who are unable to be weaned from CPB.
- Severe preoperative mitral insufficiency and ventricular dysfunction often result in postoperative hemodynamic instability and increased perioperative mortality.
Mitral regurgitation

- The optimal hemodynamic state for a patient with moderate or severe MR consists of afterload reduction, adequate preload and contractility, and high–normal HR.
- High-dose synthetic narcotics will need to be combined with a vagolytic agent such as pancuronium.
- Volatile agents are acceptable as long as they do not unduly depress contractility and they maintain HR; halothane may not be desirable.
- TEE and LAP monitoring are crucial to determine the presence of residual MR, or the occasional creation of mitral stenosis.

Pericardial effusion and tamponade

- Patients with tamponade physiology have dyspnea, tachycardia, distended neck veins, narrow pulse pressure, and pulsus paradoxus.
- Induction of general anesthesia, muscle relaxation, tracheal intubation, and positive pressure ventilation may precipitate cardiovascular collapse.
- Drainage of a small amount of the pericardial fluid with sedation (ketamine) and local anesthesia should be performed if possible.
- Etomidate is the preferred drug for induction of general anesthesia.
- Personnel and equipment for immediate drainage and resuscitation must be immediately available.
- Echocardiographic guidance is essential for closed procedures not under direct vision, e.g., pericardiocentesis.

References


61 Beck CS. Two cardiac compression triads. JAMA 1935; 104:714–16.