PDA, ASD, VSD

Reference: Anesthesia for Congenital Heart Disease
Authors: Andropoulos, D., Stayer, S. and Russell, I.
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Patent Ductus Arteriosus

The ductus arteriosus is an essential component in normal fetal circulation; it becomes functionally closed within 10-15 hours after birth, and permanently closes by thrombosis, intimal proliferation, and fibrosis in the first 2-3 weeks after birth. Functional closure is initiated by several mechanisms including aeration of the lungs, removal of prostaglandins produced in the placenta, increased arterial $P0_2$ and release of vasoactive substances (bradykinin, thromboxanes, and endogenous catecholamines).[5-7] Isolated persistent patent ductus arteriosus (PDA) occurs in approximately 1 : 2500 to 1 : 5000 live births, the incidence is higher for premature births and this defect is two to three times more common in females than in males.[5,8] PDA is also found as part of other complex congenital heart defects and is usually the source for pulmonary or systemic blood flow in patients with a functional single ventricle before palliative repair.

Anatomy

Embrylogically, the ductus arteriosus arises from the distal portion of one of the sixth paired aortic arches.[5] The PDA is a vascular communication between the descending aorta and pulmonary artery. The PDA most commonly arises from the aorta, just distal to the left subclavian artery and attaches to the left pulmonary artery (Fig. 18.2).

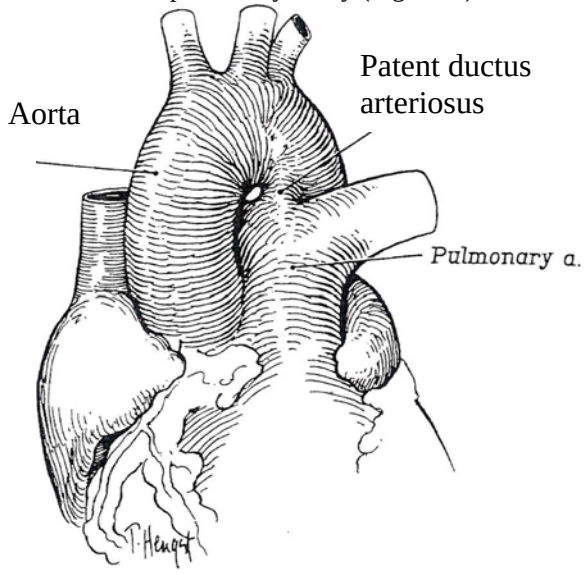

Fig. 18.2 Patent ductus arteriosus. Reproduced with permission from Cooley DA, Norman Jc. Closure of patent ductus arteriosus. In: Cooley DA, Norman Jc. *Techniques in Cardiac Surgery.* Houston, TX: Texas Medical Press, 1975: 10-17.

Pathophysiology and natural history

The degree of left-to-right shunting depends on several factors, the actual dimensions of the shunt as well as the relative ratio of *PVR* and *SVR.* The shunt dimensions of importance include the diameter and length of the PDA; shorter connections with larger diameters produce less resistance, i.e. allow greater flow. In patients with large PDAs, the diastolic runoff into the pulmonary artery results in lowered aortic diastolic pressure, which may increase the risk of myocardial ischemia, especially in the presence of anemia or lowered *SVR*..

The consequences of a PDA left untreated depend on many factors. A small PDA may be hemodynamically insignificant and unrecognized. The larger the PDA or left-to-right shunt, the more likely the progression to CHF, pulmonary hypertension, and in extreme cases, reversal of the shunt. In premature infants, PDA is associated with increased morbidity from associated respiratory distress syndrome, necrotizing entercolitis, and intracranial hemorrhage.

Surgical approaches

In newborns, surgical treatment is usually reserved for patients who fail medical treatment with indomethacin. The usual surgical options include posterolateral thoracotomy with ligation or division of the PDA or video-assisted thoracoscopic surgery (VATS). Surgical approaches have mortality approaching 0% and minimum morbidity; however, mortality rates in premature neonates is slightly higher.6 Complications of surgical treatment include, bleeding, chylothorax, vocal cord paralysis (injury to recurrent laryngeal nerve), pneumothorax, atelectasis, recurrence of patency, and inadvertent ligation of the pulmonary artery or descending aorta.[6]

Video-assisted thoracoscopic surgery has increasing popularity due to decreased pain, decreased hospital cost (secondary to decreased hospital stay), and avoidance of post-thoracotomy syndrome (rib fusion, chest wall deformities, scoliosis, and compromise of pulmonary function). Disadvantages of VATS include intraoperative desaturations and hypercarbia, as well as higher morbidity during the surgicallearning curve.[9-11]

Transcatheter closure techniques

Many catheter methods have been developed to nonsurgically close a PDA and include the Gianturco coils, the Gianturco-Grifka coil bag and the Amplatzer duct occluder (not Food and Drug Administration [FDA] approved).[8,12,13] These methods are considered safe, efficacious, and cost effective when compared to surgical closure. Risks of transcatheter approaches include arrhythmias, embolization of the device, and incomplete

closure.[12-14] In addition, there are size limitations in small infants.

Anesthetic considerations

The anesthetic management for PDA ligation depends upon factors such as patient's clinical condition, prematurity, coexisting disease, body weight, and surgical technique. Standard ASA monitors are used, along with pulse oximetry of both upper and lower extremities which will assist in detecting inadvertent ligation of the descending aorta. In addition, placing a non-invasive blood pressure cuff on both upper and lower extremites will assist in determining if the PDA ligation produced some degree of coarctation of the aorta. Large volume venous access (which may be a 22 or 24 gauge Lv. in a premature infant) and forced air-warming devices are also recommended. Among patients with coexisting disease, intra-arterial pressure monitoring provides a method of assessing arterial blood gases, electrolytes, hematocrit, and acid-base status.

Neonatal patients commonly develop hemodynamic instability from exposure to inhaled anesthetics and benefit from an intravenous anesthetic technique using opioids such as fentanyl and possibly a benzodiazepine along with muscle relaxation. Fentanyl-based anesthesia reduces the neonatal stress response and improves postoperative outcome. [15]

Neonatal PDA ligation is often performed in the newborn intensive care unit to avoid the additional risks of transport, need for ventilator changes, and hypothermic exposure. High spinal anesthesia, caudal and thoracic epidural techniques have all been described as safe and producing faster recovery.[16]

Lung isolation improves surgical exposure, especially for VATS surgical techniques, but may require ventilation with 100% inspired oxygen to maintain acceptable oxygenation. Prior to lung isolation, efforts should be used to limit the degree of left-to-right shunting by maintaining or improving pulmonary vascular tone: minimize the FIO_2 and maintain $PaCO_2$ between 40 and 50.

Atrial Septal Defects

The right and left atria are normally divided from fusion of two septa, the septum primum and the septum secundum. The septum primum develops during the fourth week and the septum secundum develops during the fifth week of gestation.[5] The septum secundum forms an incomplete partition and leaves an opening called the foramen ovale. The septum primum becomes the valve of the foramen ovale. [5] Five different types of atrial septal defects exist: (i) secundum; (ii) primum; (iii) sinus venosus;. (iv) patent foramen ovale (PFO); and (v) coronary sinus (Fig. 18.4). Isolated ASDs are more common in females than males by a factor of 2 : 1.

Atrial septal defects make up approximately 5-10% of all congenital heart defects with the secundum ASD comprising nearly 80% of ASD.6 A probe patent foramen ovale (PFO) is found in approximately 30% of normal adult hearts. Atrial septal defect may be isolated or associated with other congenital heart defects, where it may be a life saving communication allowing mixing of blood between the pulmonary and systemic circulations, such as total anomalous pulmonary venous return (TAPVR), tricuspid atresia, and transposition of the great arteries

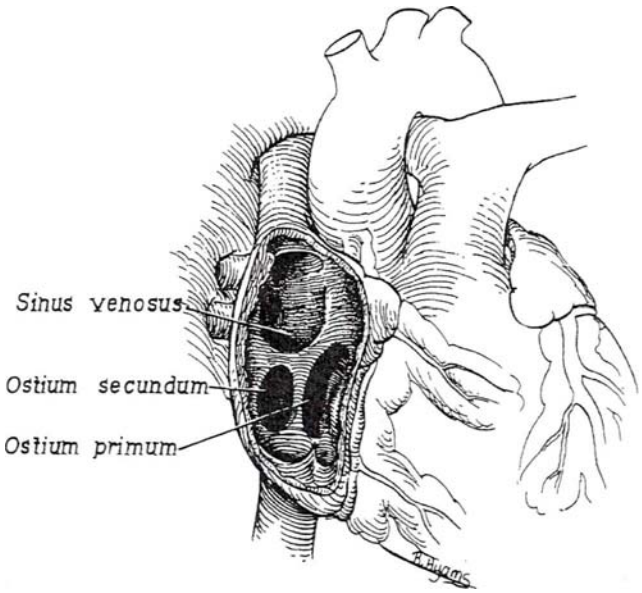

Fig. 18.4 Composite of major types of atrial septal defect. See text for explanation. Reproduced with permission from Cooley DA, Norman Jc. Closure of atrial septal defect. In: Cooley DA. Norman JC. *Techniques in Cardiac Surgery*. Houston, TX: Texas Medical Press, 1975: 70-9.

Anatomy

Secundum atrial septal defect

The secundum ASD is contained within the area bordered by the limbus of the fossa ovalis.[26] It results from an abnormal reabsorption of the septum primum or defective formation or shortening of the septum secundum. Combinations of these abnormalities may contribute to large defects.

Primum atrial septal defect

The primum ASD results from abnormalities in formation of the septum primum. It is frequently associated with atrioventricular canal (A VC) defects, especially the partial atrioventricular canal (P A VC) that includes a cleft in the anterior leaflet of the left atrioventricular valve. These A VC defects are due to defects in fusion of the endocardial cushions.

Sinus venosus atrial septal defect

Sinus venosus defects result from abnormal development of the septum secundum or the sinus venosus, the primitive venous collecting chamber. The most common type is located near the superior vena cava (SVC) orifice and is associated with partial anomalous pulmonary venous return (PAPVR) involving the right upper and middle pulmonary veins. Defects near the orifice of the inferior vena cava also exist and may involve PAPVR of the right lower pulmonary vein.

Patent foramen ovale

Patent foramen ovale results from failure of fusion of the septum primum to the limbus of the septum secundum. Patent foramen ovale is normal during fetal life as blood passes from right to left bypassing the lungs in fetal circulation. Following birth, as the *PVR* drops and *SVR* increases, the foramen ovale closes, but may not fuse.

Coronary sinus ASD

Coronary sinus ASD, also called an unroofed coronary sinus, results from an absence in the wall between the coronary sinus and the left atrium. This allows blood from the left atrium to drain into the right atrium via the coronary sinus. Persistent left SVC is also associated with this defect.[2]

Pathophysiology and natural history

The amount of left-to-right shunting at the atrial level is dependent upon two factors, the size of the defect and the relative compliance of the right and left ventricles. Shunting occurs primarily during diastole, when the atria contract and atrioventricular valves open, and produces a volume burden on the cardiovascular system that is proportionate to the amount of shunting (see the discussion in the Introduction).

Isolated ASDs are usually asymptomatic in infants and during childhood despite the increased volume load on the right ventricle. Congestive heart failure usually occurs after the second or third decade of life due to chronic right ventricular volume overload. Pulmonary hypertension can occur in up to 13% of unoperated patients youger than 10 years of age; however, progression to Eisenmenger's syndrome is unusual. The increase in right atrial size may predispose to atrial arrhythmias, and patients with a Qp : Qs of 2 : 1 or less have an 11 % incidence, whereas those with a Qp : Qs of 3 : 1 or greater have a 38% incidence of atrial arrhythmias.[27] An ASD is sometimes discovered during a neurologic work-up for transient ischemic attacks or strokes from paradoxical emboli.[2]

Surgical approaches

Surgical repair of an ASD is usually recommended between the ages of 3 and 5 years.[28] Spontaneous closure of small secundum type ASD can occur in up to 87% of infants in the first year of life, and controversy exists regarding the closure of small ASDs that are asymptomatic. Conventional surgical treatment involves median sternotomy with the use of CPB to perform a primary repair or patch closure, with surgical mortality approaching 0%. Sinus venosus defects are usually repaired using a patch to close the ASD and baffle the anomalous pulmonary veins to the left atrium. Many centers now favor minimally invasive surgery via a partial sternotomy,[29-31] because of the advantage of improved cosmetic result with similar morbidity and mortality to complete sternotomy. Postoperative dysrhythmias are reported in 23% of patients, and as many as 2% of patients may need a pacemaker following surgery.[6]

Transcatheter closure techniques

Transcatheter ASD closure in the cardiac catheterization laboratory has dramatically reduced the number of operative repairs. The CardioSEAL septal occluder (Nitinol Medical Technologies, Inc., Boston, MA) and the Amplatzer septal occluder (AGA Medical Corp, Golden Valley, MN) are the most common devices used. [13] These procedures are usually performed under general anesthesia with the use of TEE to guide placement. However, new intracardiac echocardiography using intravascular two-dimensional imaging may eliminate the need for TEE and reduce the need for general anesthesia.[32] Transcatheter closure is safe, associated with decreased hospital stay, lack of a surgical scar, avoidance of CPB, and limits the need for general anesthesia. Limitations to transcatheter closure of ASD are based on patient size large introducer sheaths), type of ASD (usually only PFO or secundum), and requires the presence of an adequate tissue rim for the device to attach.

Anesthetic considerations

Patients with ASD are generally asymptomatic, and do not have pulmonary hypertension. Therefore, the induction of anesthesia can be easily tailored' to either inhalation or intravenous technique. Whenever possible, patients should have an intraoperative TEE performed prior to incision, because transthoracic echocardiographic studies are sometimes unable to visualize all four pulmonary veins, thereby excluding the possibility of PAPVR. During surgery, TEE can be helpful to assess de-airing of the left heart and adequacy of the repair. The majority of patients have good myocardial function and do not require inotropic support perioperatively. Maintenance of anesthesia may consist of inhaled agents, intravenous agents, regional anesthesia, or a combination. Regional techniques are favored by some to assist in early extubation.[33,34] Tracheal extubation in the operating room has been shown to decrease patient charges, without compromising patient care when compared to extubation in the intensive care unit.35 Whatever technique is chosen, the primary goal for the uncomplicated ASD patient should include preparation for an early extubation either in the operating room or within the first 4 hours postoperatively.

Ventricular Septal Defects

Ventricular septal defect is the most common congenital heart defect, comprising approximately 20% of all congenital heart defects, with an incidence between 2.6 and 5.7 in 1000 live births.[36-38] Ventricular septal defect is associated with a variety of inherited conditions, including trisomy 13, 18, and 21 as well as the VACTERL (vertebral, vascular, anal, cardiac, tracheoesophageal, renal, and limb anomalies) association and CHARGE (coloboma, heart anomaly, choanal atresia, retardation, and genital and ear anomalies) syndrome.[39] Ventricular septal defects are found as isolated defects and as part of other complex congenital heart defects. Embrylogically, the primitive left ventricle is formed from the ventricular portion of the bulbis cordis and the primitive right ventricle is formed from the proximal portion at approximately 23-25 days gestation. A communication between the right and left ventricles defines a VSD and five different types of VSD exist: (i) perimembranous; (ii) subpulmonary; (iii) muscular; (iv) malalignment; and (v) canal (Fig. 18.5).

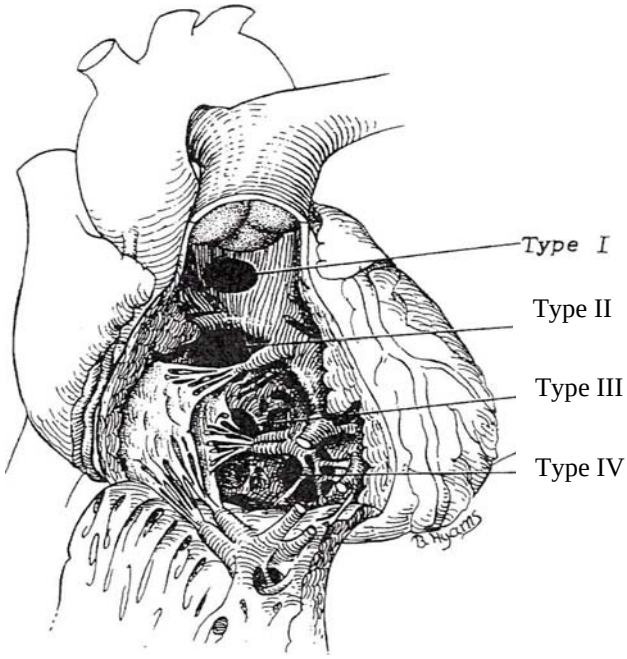

Fig. 18.5 Composite of major types of ventricular septal defect. Type I, subpulmonary; type II, perimembranous; type III, canal or inlet-type; type IV, muscular. See text for further explanation. Reproduced with permission from Cooley DA, Norman Jc. Closure of ventricular septal defect. In: Cooley DA, Norman Jc. *Techniques in Cardiac Surgery.* Houston, TX: Texas Medical Press, 1975: 80-7.

Anatomy

Perimembranous ventricular septal defect

The perimembranous VSD is a communication adjacent to a portion of the membranous septum and the fibrous trigone of the heart, where the aortic, mitral, and tricuspid valves are in fibrous continuity with the tricuspid, aortic, and mitral valves.6 These infracristal defects are the most common VSD subtype, occurring in approximately 80%.

Subpulmonary ventricular septal defect

The subpulmonary VSD is located within the outlet septum, above the crista supraventricularis and border of the semilunar valves; and comprises approximately 5% of all VSDs. As a result of the location of this defect, a Venturi effect may be produced by the jet of blood flowing through the VSD causing the right or non-coronary aortic cusp of the aortic valve to prolapse toward the defect producing aortic insufficiency.6 This type of lesion is more common in the Asian population.40

Muscular ventricular septal defect

Muscular VSDs are located within the muscular portion of the interventricular septum. These defects can be multiple and represent approximately 2-7% of VSDs.

Malalignment ventricular septal defect

Malalignment VSDs occur from malalignment of the infundibular septum and the trabecular muscular septum. These defects usually occur as a component of a more complex cardiac defect, most commonly tetralogy of Fallot.

Canal type ventricular septal defect

Canal type VSDs are located in the posterior region of the septum beneath the septal leaflet of the tricuspid valve.[32] These inlet defects accounts for approximately 10% of VSDs.

Pathophysiology and natural history

Isolated VSDs produce left-to-right shunting at the ventricular level, predominantly during systole. The term restrictive VSD refers to a limitation in the amount of flow across the defect based on size; and in this case a pressure gradient exists between the left and right ventricles. An unrestrictive VSD has flow limited only from the relative pulmonary to *SVR;* and therefore, no pressure gradient exists between the left and right ventricles. Fifteen percent of patients with large VSDs develop pulmonary hypertension which will progress to the development of pulmonary vascular obstructive disease by the age of 20 years.[41]

Symptoms range from asymptomatic to signs and symptoms of CHF. The rate and degree of progression of symptomatology depends on the patient age, size of the defect, and the degree of left-to-right shunting. Infants who have unrestrictive VSDs develop symptoms of CHF in the first 3 months of life because of the physiologic decline in *PVR.* Spontaneous closure of small perimembranous and muscular VSDs occurs in as many as 50% of patients,[41-43] and such patients are typically asymptomatic.

Surgical approaches

Surgical repair of VSD usually involves patch closure or occasionally primary closure using CPB via median sternotomy. Perimembranous and canal type VSDs are most commonly repaired via a right atriotomy, which may require detachment of the septal leaflet of the tricuspid valve for exposure. Subpulmonary VSDs are most commonly repaired via the transpulmonary approach. Midmuscular VSDs are most commonly repaired via right atriotomy, and anterior or apical muscular VSD may be approached using right ventriculotomy. However the use of right ventriculotomy carries the risks of conduction disturbances and ventricular dysfunction later in life. At many institutions, symptomatic patients with lesions that are not approachable via right atriotomy are usually treated with pulmonary artery banding until the patient is larger allowing transatrial repair. Pulmonary artery banding is also utilized for multiple muscular VSDs and in patients that are high-risk candidates for CPB. Partial median sternotomies as well as small right anterolateral thoracotomies are advocated by some because of improved cosmetic results.[44,45] Video-assisted cardioscopy (VAC) is used in some centers to improve visualization of small intracardiac structures in limited spaces during open heart surgery for congenital heart repairs.[31,46-48] Videoassisted cardioscopy has been successfully utilized for a variety of intracardiac repairs including ASD, VSD, tetralogy of Fallot, double outlet right ventricle (DORV) and AVC.

Timing for surgical repair varies depending on age, signs, and symptoms. Patients less than 6 months of age are repaired if they manifest uncontrollable CHF and failure to thrive. Patients between 6 and 24 months of age undergo repair to treat CHF symptoms or pulmonary hypertension. Patients older than 24 months undergo repair for Qp : Qs greater than 2 : 1. Among patients with subpulmonary VSD, the presence of aortic insufficiency is an indication for surgical repair to prevent further progression of the valvular insufficiency,[49.50] A defect size greater than 5 mm is repaired to avoid progression to aortic cusp prolapse and aortic insufficiency, and defects less than 5 mm can be managed conservatively.[40]

Mortality for uncomplicated VSD in older patients is less than 1-2%.[51] Mortality for VSD repair in infants during the first year of life is less than 5%.[52]

Transcatheter closure techniques

Transcatheter closure of muscular VSDs has been performed successfully.[53-56] The use of the CardioSEAL septal occluder (Nitinol Medical Technologies, Inc., Boston, MA) is approved for use in the USA, and the Amplatzer septal occluder (AGA Medical Corp., Golden Valley, MN) is undergoing clinical trials in the USA. Indications for use of the devices include all types of muscular VSDs, including apical and multiple. Transcatheter techniques may serve as an adjunct to surgery or an alternative to surgery in selected patients. Intraoperative use of VSD closure devices during CPB for defects with difficult surgical closure has been described,[57] and postoperative residual VSDs or "Swiss cheese" type muscular VSD may be preferentially treated by this technique. The major limitation in the application of this technique is related to the size of the sheaths necessary for device delivery, precluding use in infancy. Complications of device closure include need for blood transfusion, tricuspid valve regurgitation, and device embolization.[53,55]

Anesthetic considerations

Anesthetic management for the patient with VSD is similar to that of ASD. Pulmonary hypertension may develop early, especially in patients with trisomy 21, and preoperative chest radiograph revealing decreased pulmonary vascular markings is indicative of pulmonary hypertension.[58-61] Such patients may respond to the use of inhaled NO prior to termination of CPB and/or in the postoperative period. Right heart failure with decreased CO may result if pulmonary hypertension is not controlled, and is improved from the use of dopamine, milrinone, dobutamine, or isoproterenol.

Conduction disturbances, particularly atrial-ventricular heart block may be transient or permanent and is reported to occur in up to 10% of patients post- VSD repair;[6] however, the experience at Texas Children's Hospital is that less than 1 % of patients require permanent pacing after VSD closure. If heart block develops, treatment with atrioventricular synchronous pacing using temporary pacing wires is indicated. Junctional ectopic tachycardia is sometimes observed in patients less than 1 year of age after repair for lesions that involve VSD repair, most commonly after tetralogy of Fallot repair. Treatment includes cooling to 35°C, increased anesthetic depth, paralysis, procainamide, esmolol, or amiodarone.[6]

Intraoperative use of TEE will help recognize residual VSDs, intra cardiac air, and assess ventricular volume and function. Small muscular VSDs will become apparent after closure of larger VSDs. Frequently these smaller defects, especially if near the apex, may not be amenable to surgical repair or worth the risk of returning to CPB.

Patients with uncomplicated VSDs are good candidates for extubation in the operating room or early after arrival in the intensive care unit.