Patent ductus arteriosus is a common congenital heart disease that can be present isolated but is usually present with other co-existing congenital heart anomalies. Patients usually have a previous history of hypoxia, or premature baby or have a maternal history of rubella infection. Echocardiography is the diagnostic modality of choice for the confirmation of the diagnosis. The administration of indomethacin can stimulate the closure of the patent ductus arteriosus. Surgical treatment should be reserved for highly symptomatic patients with a large patent ductus arteriosus.

Definition of Patent Ductus Arteriosus (PDA)

Humans need blood that must pass through the lungs to survive; it is called oxygenated blood. However, while still in the womb, it is not necessary for a fetus’ blood to pass through the lungs for it to become oxygenated because Oxygen supply is maintained by maternal blood. In place of lungs, a fetus has a ductus arteriosus which links the aorta to the pulmonary artery, enabling blood supply to avoid circulation through the lungs. Instead, blood travels directly from the pulmonary artery into the aorta and is circulated.
Naturally, immediately after birth, the ductus arteriosus is supposed to close so that oxygenated blood can be supplied by lungs through the air. In a few cases, the ductus arteriosus fails to close (or patent), leaving an open hole in the heart. This is referred to as Patent Ductus Arteriosus (PDA).

Epidemiology of Patent Ductus Arteriosus (PDA)

Patent ductus arteriosus has an estimated incidence of 2 in 10,000 live births in the United States. The incidence of PDA is higher in premature infants and can reach up to 60%. PDA represents 5-10% of all congenital heart diseases.

The most common risk factors for PDA are a history of perinatal asphyxia, birth at high altitude, the presence of other congenital heart diseases in the child, a positive family history of PDA, and low birth weight. Maternal infection with rubella might be associated with a PDA.

The incidence of PDA is believed to be equal in people of different races. Female to male ratio of isolated PDA is 2:1.

PDA is present since birth, it is known to eventually close in children. With a history of asphyxia, and it can be an incidental finding on a routine chest examination of a child. During heart auscultation, a continuous murmur might be heard and the diagnosis of PDA can be suspected.

Isolated PDA has been associated with excellent prognosis especially in full-term babies. Spontaneous closure of PDA has been reported in up to 75% of infants within the first three months of life. Patients who do not undergo spontaneous closure within three months are unlikely to get closed. A large PDA can cause pulmonary hypertension and congestive heart failure if left untreated.

Etiology of Patent Ductus Arteriosus (PDA)

The etiological factors of PDA can be assessed as follows:

- The exact etiology of PDA is unknown but familial cases have been reported. Genetic causes are not established.
- Some possible risk factors for PDA are maternal rubella infection during the first trimester of pregnancy, maternal alcohol use, and maternal use of the antiepileptic phenytoin drug.
- Prematurity has been also associated with an increased risk of isolated PDA.
- Patients with hypoxia due to asphyxia, coexisting congenital heart disease, offsprings of those living in high altitudes, and anemia is more likely to have a persistent PDA.

Classification of Patent Ductus Arteriosus (PDA)

According to Krichenko, PDA can be classified based on computed tomography angiography, as follows:

- Type A: it has a cone-shaped ductus, along with a protruding aortic ampulla which becomes narrow towards the end of the pulmonary artery.
- Type B: It is small, wide and blends with the pulmonary artery.
Type C: The ductus is shaped like a tube and is long, without constrictions
Type D: The ductus is convoluted, with several constrictions.
Type E: It is drawn out with remote constriction

Premature children may have a tortuous ductus which is not included in the above classification. This type, usually called type F, is bigger, longer and narrows with a tortuous appearance at the pulmonary artery end.

Pathophysiology of Patent Ductus Arteriosus (PDA)

Patients born with a PDA might have pulmonary over-circulation. The larger diameter of the PDA and a shorter length of the communication is associated with larger left-to-right shunting of blood. The blood starts flowing through the PDA, goes to the pulmonary arteries and lungs, returns to the left atrium via the pulmonary veins and finally goes to the left ventricle. The blood is then pumped from the left ventricle to the aorta, and part of it is again shunted through the PDA.

Because of this shunting and volume overload to the left atrium and ventricle, it is understandable why patients with prolonged periods of time with untreated PDA might develop ventricular dilatation. Another important pathological change that is observed with PDA is pulmonary hypertension which is due to pulmonary over-circulation and unrestricted blood flow through a large PDA.

Low oxygen tension in the blood has been linked to increased production of prostaglandin E by the ductus arteriosus wall and persistence of the patency of the ductus arteriosus. Prematurity elevates the risk of PDA because the metabolism of prostaglandins by the lungs is not yet fully developed. Cyclooxygenase (COX-2) is overexpressed in premature infants. This enzyme is responsible for the production of prostaglandin E.
Clinical features of Patent Ductus Arteriosus (PDA)

Symptoms

A lot of patients who are not diagnosed with PDA at birth complain of a heart murmur without other symptoms, while others are discovered accidentally while an echocardiogram is being performed for another purpose entirely. A few patients may complain of exercise intolerance or reactive airways disease may have been detected. Several patients with asymptomatic PDA in childhood develop congestive heart failure later in life due to chronic volume overload, with symptoms of atrial fibrillation at the beginning.

Furthermore, symptoms of a PDA range from mild to severe, depending on the size of the opening. A small hole may present no symptoms, a heart murmur usually points to a PDA. Symptoms of PDA in a child include:

- Excessive Sweating
- Breathlessness
- Labored breathing
- Getting tired easily
- Poor weight gain
- Lack of interest in food

An adult with an undetected PDA will likely experience the following symptoms:

- Palpitations in the heart
- Shortness of breath

Signs

The characteristic sign of a PDA is a continuous murmur at the boundary of the upper left sternum.

Sometimes, patients with moderate or large holes have a diastolic rumble at the apex of the heart. Furthermore, there is an obvious impulse on the left ventricle along with an increased pulse pressure, which is more obvious on the periphery. It is very rare to notice rales even if the shunt is large, except if the patient is old and has congestive heart failure.

Cyanosis is observed in individuals with Eisenmenger's syndrome, and some may have differential cyanosis (in which there are cyanosis and clubbing of the toes, without involving the fingers). Cyanosis is likely to be greater with reduced systemic vascular resistance, especially when temperatures are high and after exercise.

With small holes, a systolic and diastolic murmur may be absent; instead, there may be a high-frequency decreasing murmur during diastole resulting from pulmonary regurgitation and/or a murmur that is holosystolic in nature resulting from regurgitation in the tricuspid valve.

If the individual has a dysfunction of the right ventricle, peripheral edema may be present later with the progress of the disease.
Investigations of Patent Ductus Arteriosus (PDA)

Chest Radiograph

A chest radiograph may appear normal or reveal an enlarged heart, subject to the volume of ductal shunting, with an enhanced outline of pulmonary vessels. The ductus may appear calcified in older people.

Electrocardiogram

The ECG may reveal sinus tachycardia, atrial fibrillation, left ventricular hypertrophy, as well as a left atrial enlargement in individuals with medium to large holes. It may appear totally normal in patients with smaller holes. An enlarged right atrium, hypertrophy of both ventricles and increased pressure of the pulmonary artery is observed in patients with large holes.

Echocardiogram

An echocardiogram is the best procedure to authenticate the diagnosis and characteristics of a PDA. The echocardiogram is helpful in categorizing a PDA as silent, minor, medium or large as well as identifying any other related heart defects. To correctly measure the dimension of the cardiac chambers and quantify the function of the left ventricle, M-mode echocardiography is utilized. The sizes of each chamber remain normal with small ductus arteriosus, even though the left atrium and ventricle may be enlarged.

Color Doppler can appraise the extent of ductal shunting, no matter the size of the hole. However, in individuals with high resistance at pulmonary vessels and faint right-to-left flow speed, it may be hard to recognize a ductus arteriosus no matter the size. Diastolic pressure from the pulmonary artery can be appraised by measuring the Doppler speed of pulmonary regurgitation.
Magnetic Resonance Imaging and Computed Tomography

In an adult where surgical intervention is being contemplated, computed tomography can evaluate the extent of calcification. For individuals whose PDA has an abnormal shape, magnetic resonance imaging and computed tomography can be used to observe the outline of the PDA.

Phonocardiogram
Cardiac Catheterization

Catheterization can be used to assess the extent of shunting and degree of pulmonary vascular resistance. It can also be used to evaluate if the closure of the ductus is a viable option.

Differential diagnosis of Patent Ductus Arteriosus (PDA)

First of all, it is important to differentiate between a clinically important and non-significant PDA. A medically important PDA is typified by respiratory issues including labored breathing, breathlessness, and metabolic acidosis.

<table>
<thead>
<tr>
<th>Venous hum</th>
<th>The murmur is more pronounced on the right side of the heart but reduces as the patient gets into a supine position along with local compression. If the evidence is not sufficient, an echo is carried out which will come out normal.</th>
</tr>
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<tbody>
<tr>
<td>Coronary artery fistula</td>
<td>With a coronary artery fistula, the murmur is continuous and is prominent in the lower part of the precordium. On an echocardiogram, a coronary fistula will show as a continuous flow of blood into the right ventricle. Its location is also different from that of a PDA.</td>
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Left-sided shunts
Examples of left-sided shunts include ventricular septal defect and atrioventricular septal defect. While the presentations may be similar on an ECG and CXR, the murmur of the majority of left-to-right shunts is noticed only during systole. However, in case of a big connection between the aorta and pulmonary artery, the diagnostic results will be the same as those of a large PDA. An echocardiogram will differentiate between them based on their typical appearance and position.

Aortic regurgitation
Individuals with this condition are diagnosed at older ages. A symptom of getting tired easily with exercise is present, even though they may not display tachypnoea. Typically, a shrill murmur that reduces during diastole is heard at the boundary of the lower-left sternum. An echocardiogram scan can easily differentiate between an aortic regurgitation and a PDA.

### Management of Patent Ductus Arteriosus (PDA)

The only treatment for a child with an asymptomatic PDA is prophylactic antibiotic therapy before dental procedures or instrumentation.

The administration of non-steroidal anti-inflammatory drugs such as indomethacin can stimulate the closure of a PDA. The frequency and duration of non-steroidal anti-inflammatory drugs administration should be completely based on the level of brain natriuretic peptide in the serum.

Digoxin and diuretics are indicated in the treatment of patients with congestive heart failure due to a PDA until the patients are old enough to undergo ligation of the PDA.

Children older than one-year-old who have a large and symptomatic PDA should undergo catheter-based closure as spontaneous closure after one year is very unlikely.

Different methods of catheter-based closure exist and include Gianturco spring occluding coils, Amplatzer duct occluder, Rashkin ductus occlusion device, and ligation of the PDA.

**Patients with a large PDA who do not respond to indomethacin, have congestive heart failure, or recurrent subacute bacterial endocarditis should undergo surgical ligation of the PDA.**

Patients with aortic atresia, hypoplastic left ventricle with mitral valve atresia, pulmonary atresia, tricuspid atresia and transposition of the great arteries should not undergo closure of their PDA until correction of the pre-existing congenital heart condition is complete. Patients with severe pulmonary hypertension are not good candidates for surgical closure of their PDA because they are unlikely to have a significant decrease in their pulmonary arterial pressure after surgery.

**Pharmacological treatment**

Treatment with diuretics and digoxin is beneficial in patients who do not show symptoms of PDA. Angiotensin-converting enzymes are used to reduce after-load, even though studies confirming these claims are unavailable. Individuals with atrial fibrillation are likely to benefit from antidysrhythmic and anticoagulation drugs. All patients with PDA, including asymptomatic PDA, benefit from a regimen of infective endocarditis treatment for at least 6 months after surgical closure.

Medications should be prescribed for individuals with congestive heart failure resulting from PDA until a surgical or transcatheter procedure to close the hole is carried out. However, if symptoms persist or the patient is diagnosed with cardiomegaly, drug therapy should be continued.

Where surgery for closure is contraindicated, PDA patients can be treated with pulmonary
vasodilators such as oxygen, PGI$_2$, calcium channel blockers, endothelin antagonists, and phosphodiesterase type V inhibitors. It should be noted that results validating these claims are anecdotal at this time.

Surgical treatment

**Indications for surgical closure of PDA include:**

Children or adult with symptoms of left-to-right shunting. For those without symptoms, closing the ductus reduces the risk of future problems. Good outcomes have been reported in adults who underwent PDA closure because removing the hole decreases pressure on the pulmonary artery.

A lung biopsy is endorsed for a patient whose pulmonary vascular resistance is greater than 8 U/m$^2$ to evaluate if the individual can undergo surgical closure. In cases where the resistance is greater than 8 U/m$^2$, the ductal closure can lead to increased pulmonary artery pressure, reduced cardiac output, and failure of the right ventricle. However, a lung biopsy may not be enough to predict a patient’s risk of undergoing a closure.

There is no indication for closing minor or asymptomatic shunts, especially in adults. After considering all the above conditions, it is very practical to carry out shunt closure in children and younger adults as they are efficient, and associated with decreased morbidity and mortality rate.

**Transcatheter Closure**

It is a high-quality treatment of PDA in children and adults, and serious complications are a rare occurrence. It is more advantageous than surgical closure where a patient’s ductus arteriosus is calcified and pulmonary vascular resistance surges.

Another advantage of this technique is that it can be done using different devices and techniques to take care of the variations in the shape and size of each PDA. Complications include distortion of flow in the proximal part of the left pulmonary artery, hemolysis resulting from high-speed residual shunting, vein thrombosis, and infection.

**Surgical Therapy**

While surgical management results in more pain and morbidity than transcatheter closure, they are extremely safe and successful especially for bigger ductus. Mortality rates are very low and lots of complete closures have been reported. Significant short term complications include bleeding, a pneumothorax, infection, etc. and long term complications can be narrowing of the aorta, incomplete closure and reopening of the PDA. However, such complications are very rare.

**References**


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