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# Cardiac Catheterization in Congenital Heart Disease

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**Abstract**

Interventional pediatric cardiology is a specialty of pediatric cardiology that deals specifically with the catheter-based treatment of congenital heart diseases. Cardiac catheterization involves the evaluation and manipulation of the heart and surrounding vessels through catheters place in peripheral vessels. In this chapter we begin by discussing the significant difference between adult and pediatric interventional cardiology. We will discuss basic hemodynamic measurements performed in cardiac catheterization and its application to congenital heart disease. Stent and balloon catheters are briefly discussed. Finally, specific catheter based interventional techniques, indications, and complications for various pediatric congenital heart disease is described.

**Keywords:** pediatric interventional cardiology, valvuloplasty, angioplasty, balloon catheter, stent

## 1. Introduction to cardiac catheterization in congenital heart disease

Cardiac catheterization in the pediatric population has similarities with catheterization in adults but very distinct differences. In adults, the primary pathology is isolated coronary atherosclerotic disease, which is exceedingly rare in pediatrics, and valvular disease. The indications, techniques, and interventions performed in pediatrics are different. There are a wide range of therapeutic procedures performed in the pediatric cardiac catheterization lab including device closure of septal defects, balloon angioplasty of stenotic lesions and valvuloplasty of stenotic valves, stenting for vascular stenosis, embolization and device closure of vessels, and even percutaneous pulmonary valve implantation. A complete assessment of the patient is important as well as evaluating and determining the best sedation, vascular access, and potential interventions.

At its most basic, cardiac catheterization is the evaluation and manipulation of the heart and related vessels by catheters placed through peripheral vessels. Most commonly, access is now obtained by the femoral artery and vein using a modified Seldinger technique, which involves placing a sheath and catheter over a wire. Alternative sites include the carotid and jugular, axillary, radial, subclavian, transhepatic, and even umbilical vessels in neonates. Anytime a catheter, wire, or sheath sits in a blood vessel, primarily an artery, there is a risk of occlusion, thromboembolism, and stroke. To prevent this, heparin is routinely administered throughout a catheterization case. Knowledge of basic technique and case-specific complications will help the practitioner in the management of these patients.

## 2. Basic hemodynamics

Traditionally, cardiac catheterization was the primary tool in the diagnosis and evaluation of congenital heart disease. Before the emergence of echocardiograms, clinical suspicion of congenital heart disease required a cardiac catheterization for definitive diagnosis, possible intervention, and pre-surgical planning. Cardiac catheterization can provide information such as angiographic images by fluoroscopic imaging during contrast injection, pressure measurements, oxygen blood saturations, and estimations of cardiac output and pulmonary vascular resistance. Pressure measurements are taken by way of a fluid-filled pressure transducer attached to a catheter with end-holes that are passed through various vessels and structures in the heart (**Figure 1**). The transducer produces cardiac pressure waveforms that are interpreted by the interventionalist (**Figure 2**). This is especially important in the evaluation of stenosis, diastolic dysfunction, and pulmonary vascular resistance.
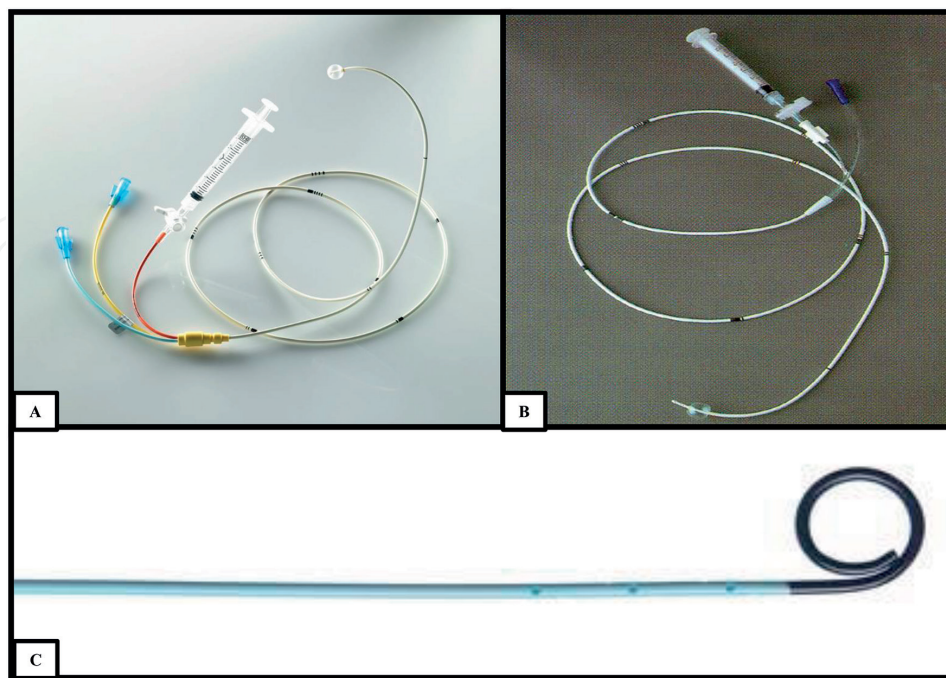


**Figure 1.** Various cardiac catheters. (A) A thermodilution catheter used to calculate cardiac output. (B) A Berman catheter with an inflatable balloon and side holes is a multiuse catheter. (C) A pigtail multiuse catheter. The multiple side holes allow for angiography of arterial vessels.

In each chamber of the heart, blood can be aspirated through a catheter and a blood oxygen saturation measured. These data can then be used to estimate a patient's cardiac output and ratio of pulmonary blood flow to systemic blood flow (Qp:Qs). In 1870, Adolph Fick described a method for calculating blood flow which was refined and is now known as the Fick Method [1]. Basically, he explained that the release of a substance (in this case, oxygen consumption) in an organ is the product of blood flow across that organ and the concentration difference of a substance (oxygen content difference) proximal and distal to the organ. In other words:

$$\text{Cardiac output} = \frac{\text{Oxygen consumption}}{\text{Oxygen content difference}} \tag{1}$$

Oxygen consumption is often estimated based on normal values in older children and adults. Oxygen content difference is primarily the product of the change in hemoglobin saturation and the oxygen carrying capacity of hemoglobin (1.36).

$$\text{Oxygen content difference} = 1.36 \times \text{hemoglobin} \times \text{difference in oxygen saturations} \tag{2}$$

Using this principle, one can estimate the blood flow through any organ by knowing the oxygen blood saturations proximal and distal to the organ and the hemoglobin concentration of blood. In this way, sampling blood in the systemic veins proximal to the heart and blood in the aorta, distal to the heart, can estimate cardiac output. Similarly, sampling blood in the pulmonary arteries and pulmonary veins can estimate pulmonary blood flow.
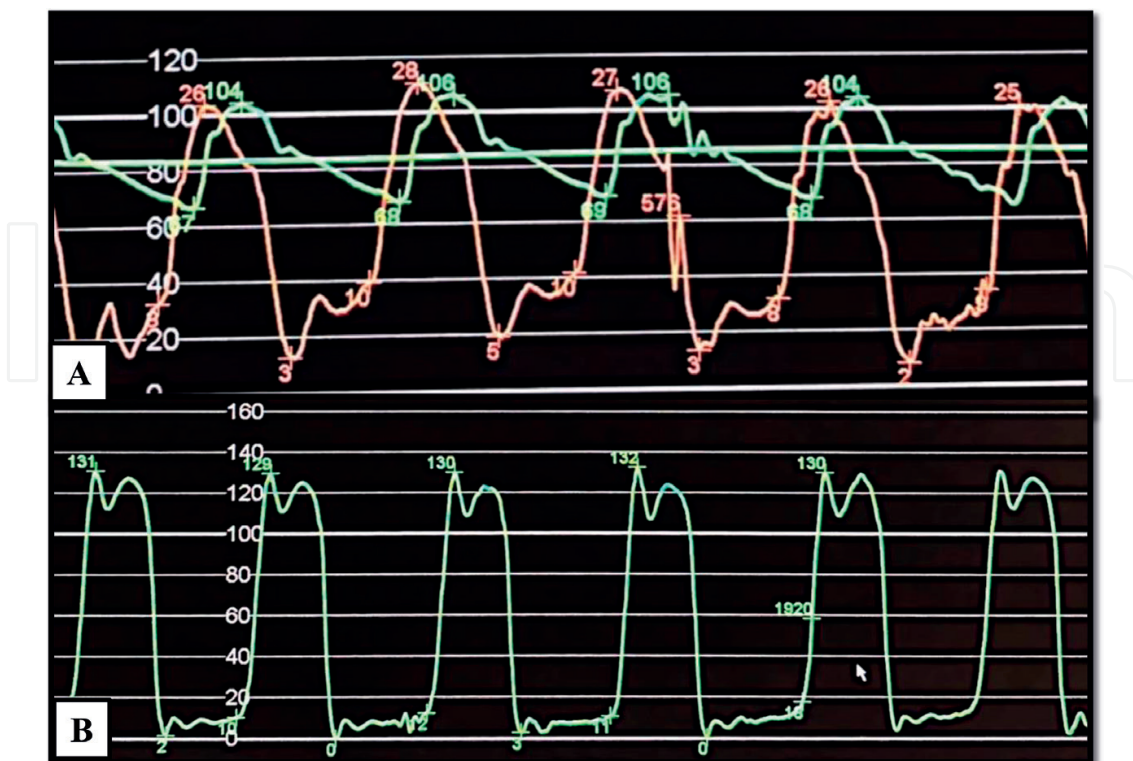


**Figure 2.** Normal intracardiac pressure tracings in a 10 year old patient. (A) A simultaneous pressure tracing of the right ventricle (red) and the femoral artery (green). (B) Pressure tracing of the left ventricle.

Resistance calculations are then based on Ohm's law of electromagnetism. Georg Ohm was a German born scientist who in 1827 described that the electric current through a conductor is directly proportional to the potential voltage difference divided by the resistance. Modified for body fluid dynamics and a quick variation, it simply states that:

$$\text{Resistance} = \frac{\Delta \text{ Pressure}}{\text{Flow}} \tag{3}$$

Ohm's law is used to calculate the pulmonary vascular resistance (PVR). The change in pressure across the lungs is estimated by the difference between the left atrial pressure and pulmonary artery pressure in a normal heart. The pulmonary blood flow is estimated by the Fick principle. PVR is an important measure in the diagnosis and management of pulmonary hypertension and pre-surgical planning.

## 3. Basic interventions

The majority of pediatric cardiac catheterization interventions include balloons, stents, and various devices. The equipment and tools of an interventional cardiologist varies based on experience level and new technological innovations. Attached to a specialized balloon catheter, an inflatable and deflectable balloon is essential in the treatment of stenosis and the deployment of stents. There are a variety of different types of balloons that range in size, shape, and compliance. These specialized balloon catheters have end holes to inflate balloons with contrast solution so they are easily visible on fluoroscopy. Using a specialized device known as a gauge, the balloons are inflated, deflated, and removed from the patient. This is useful in the treatment of vessel stenosis and valve dilation. Balloon interventions carry lesion specific risks described in the subsequent sections of this chapter.

The first intravascular stent use in children was first described in the late 1980s. Indications include those stenotic lesions that are unresponsive to balloon dilation or recur frequently. Additionally, covered stents are using in treating significant tears or aneurysms. Each stent has its own set of characteristics including their size, strength, and "shortening" ability (**Figure 3**). Stents are metal or plastic mesh tubes that are loaded over balloon catheters and expandable
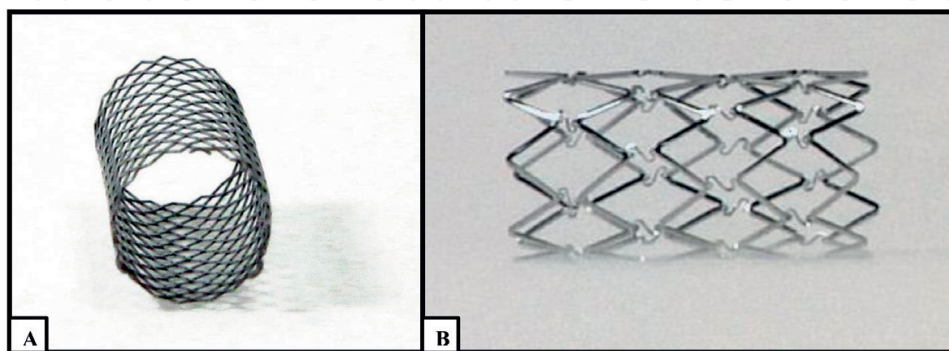


**Figure 3.** A and B are two different kinds of bare metal stents used in pediatrics. Notice the interconnected mesh wiring and "tube" like shape.

inside a vessel. Covered stents have a surgical fabric-coating that creates a contained tube that is expandable similar to bare stents. Finally, there are a number of devices used to embolize or close vessels and treat septal defects that are described in the following respective sections.

# 4. Patent ductus arteriosus

## 4.1. Introduction

The ductus arteriosus is an embryological vessel that connects the systemic and pulmonary circulation and serves to bypass the neonatal lungs. The vessel connects the pulmonary artery to the aortic arch. The patent ductus arteriosus (PDA) is a persistence of the ductus arteriosus after birth. The PDA is useful in cyanotic congenital heart diseases and is kept open until surgical palliation in many cases. This is described further in the chapter on cyanotic congenital heart disease. In the absence of a cyanotic heart lesion, the isolated PDA allows for a portion of oxygenated blood from the left side of the heart to flow back towards the lungs. Isolated PDA accounts for 10% of all congenital heart disease and is unlikely to close naturally in term infants [2]. Long term complications can include congestive heart failure, infective endocarditis, and pulmonary hypertension.

The first closure of the PDA was done by Gross and Hubbard in 1939. The first transcatheter closure of the PDA was performed by Portsmann et al. in 1967 and since then, there have been many developments in the closure of the PDA. Until recently, transcatheter closure of the PDA was not performed in infants less than 5–10 kg, but is now performed in infants as small as 600 g in some centers. Further innovation and experience in this procedure will result in a non-invasive, effective alternative to surgical PDA ligation in nearly all patients.

## 4.2. Indications

In older patients, the indications for closure of the PDA are well defined. Patients who have signs of left ventricular overload due to the left to right shunting of the PDA should be evaluated for cardiac catheterization closure. A long-standing PDA can lead to congestive left sided heart failure and Eisenmenger's syndrome similar to a ventricular septal defect. The indications for closure in neonates is more controversial. There is currently inadequate data for or against the closure of the PDA in young infants [3], but a general consensus is that transcatheter closure of the PDA in symptomatic infants at 2–3 weeks of age is both safe and effective. Alternatives include surgical ligation and medical closure with a non-steroidal anti-inflammatory drug such as indomethacin or acetaminophen. However, pharmacological treatment of the PDA is effective primarily in premature infants, and has an effective closure rate of around 68% [4].

## 4.3. Catheterization

Although the procedure is sometimes performed under only echocardiographic guidance and occasionally in the neonatal intensive care unit, we recommend fluoroscopy guidance in the cardiac catheterization lab. Traditionally, both femoral venous and arterial access is
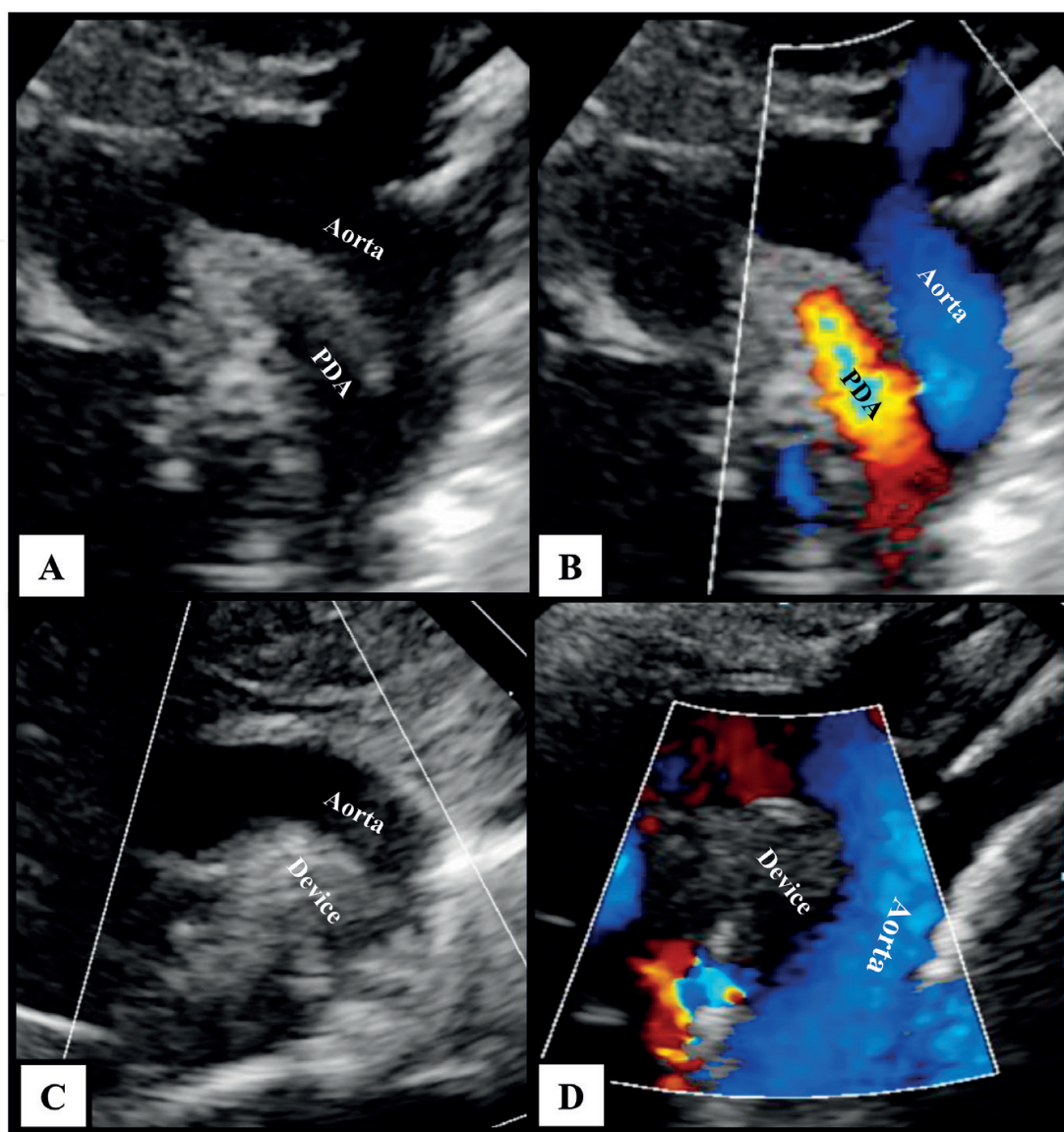
**Figure 4.** Echocardiographic images of a PDA closure in a 4 week old premature infant who could not be extubated due to pulmonary edema. (A) Two-dimensional (2D) image of the aorta and the PDA as marked. (B) Color Doppler of the aorta and PDA showing opposite flow in each vessel. There is left to right shunting in the PDA. (C and D) After closure with a device, the PDA is no longer visualized by 2D or color Doppler.

obtained. Arterial access is used as a guide for the distal end of the ductus. Recent technique innovation requires only femoral venous access and placement of an esophageal temperature probe. The probe acts as a landmark for the distal end of the ductus. The ductus is engaged by traversing a catheter through the pulmonary artery, ductus, and into the descending aorta.

First, an angiogram is performed to obtain measurements and describe the morphology of the PDA. A PDA with a pulmonary end diameter of less than 2.5 mm can occasionally be closed by coil embolization. These are spring wire coils enmeshed with polyester fibers and delivered through a catheter. The coil occludes the vessel by creating a wire and fabric framework that clots. In most PDAs, an occlusion device is used. A guidewire is used as an anchor and an occlusion device is loaded through a catheter into the PDA. There are various devices used

for PDA closure including vascular plugs and two-disk duct occluders similar to atrial septal device occluders. The diameter of the device should be at least the measured diameter of the ductus. Ideally, the device is situated entirely in the ductus arteriosus with no protrusion in either the pulmonary artery or the aorta. Once the device is in position, echocardiography is used to verify no residual shunting, and no obstruction to either the descending aorta or the branch pulmonary arteries. Once this is verified, the device is deployed (**Figures 4–6**).

## 4.4. Complications

The major complications of the procedure acutely are obstruction of the aorta or branch pulmonary arteries and embolization of the device. The rate of embolization ranges from 1 to 3% and is nearly always retrievable by catheterization without surgical intervention. Loss of distal pulses and vascular injury has been shown to occur in around 20% of premature infants who
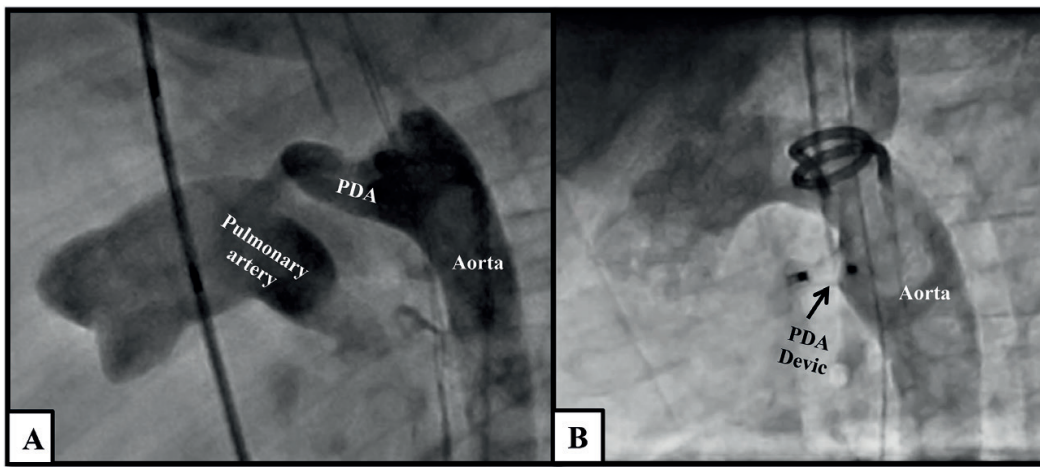


**Figure 5.** Angiographic images of a PDA closure in a 4 year old patient who presented with left atrial and left ventricular dilation. (A) Contrast injection of the PDA through a pigtail catheter located in the aorta through a retrograde arterial course. (B) Contrast injection angiogram after closure of the duct with visualization of the PDA device. There is no visible stenosis of the aorta.
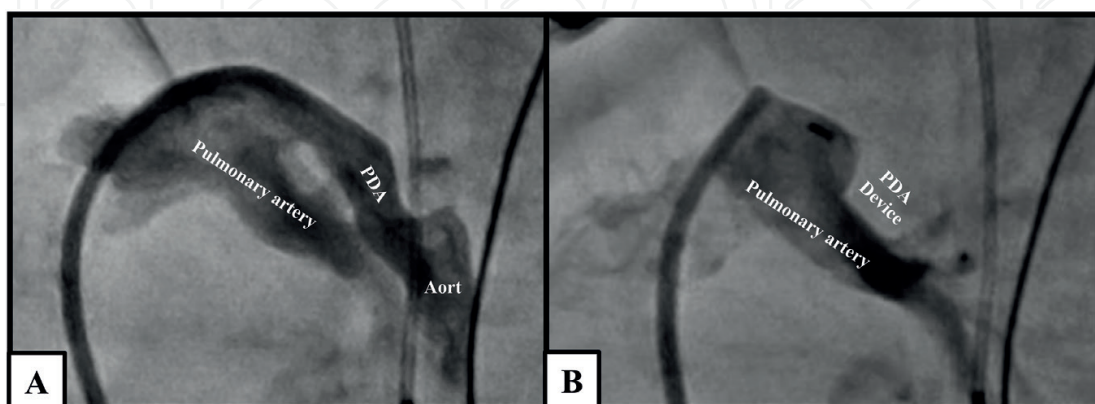


**Figure 6.** Angiograms of a PDA closure in an 800 g 28 week gestation infant who was unable to wean from ventilatory support. There was no arterial access in this patient. (A) An angiogram performed shows a large PDA similar in morphology to most premature infant ducts. (B) After closure with a microvascular plug device, there is no residual flow through the duct and the left pulmonary artery is unobstructed.

had arterial access [5]. Infants also suffer an entity known as post ligation cardiac syndrome. Six to twelve hours after ligation, these patients suffer a transient low cardiac output state occasionally requiring additional inotropic support. Reported rates are as high as 50% [6].

## 5. Atrial septal defect

### 5.1. Introduction

Atrial septal defects (ASD) are holes between the right and left atrium of the heart. Their presence is noted in nearly half of all congenital defects and is important for preserved cardiac output and adequate mixing in a variety of critical congenital heart disease as described in the section on atrial septostomy [7]. Ostium secundum defects are the result of an embryological defect in the septum primum. They are the most common type of ASD with nearly 75% fitting their description. Other types of ASDs include sinus venosus, coronary sinus, or ostium primum defects. Isolated atrial septal defects are typically asymptomatic in infants as the shunting across them is insignificant in early life. Eventually, patients may become symptomatic with frequent respiratory infections, though most are diagnosed due to the presence of a persistent murmur heard by a primary care practitioner and subsequent echocardiogram. Right ventricular and atrial dilation may be present along with a classic fixed split S2 heart sound on auscultation [8].

The first successful surgical closure of an ASD was performed in 1949. It was not until 1972 that the first transcatheter closure of ASDs were performed in animal studies [9]. The first human patient to undergo transcatheter device closure of an ASD was a 17 year old girl in 1975 using a Rashkind foam-covered, six-ribbed device [10]. Since that time, there have been numerous occlusion device revisions and innovations. Since the early 2000s, overall results from transcatheter atrial septal defect device closure have achieved results comparable if not exceeded those of surgical closure [11].

### 5.2. Indications

As stated before, patients with atrial septal defects rarely have clinically relevant symptoms. Ostium secundum defects are the only type of ASD amenable to device closure. In general, atrial septal defects should be closed in patients with right cardiac chamber enlargement with or without symptoms, a paradoxical embolism, or exercise related cyanosis. Patients who have severe pulmonary arterial hypertension unresponsive to vasodilator therapy, intracardiac thrombus, or a contraindication to antiplatelet agents should not undergo transcatheter closure. Additionally, defects must measure 36 mm or less, have adequate rims, and be located a safe distance from adjacent structures. Those that do not meet these criteria should undergo evaluation for surgical closure.

### 5.3. Catheterization procedure

All patients should have standard catheterization precautions with general anesthesia, antibiotic prophylaxis, and anti-coagulation with heparin. In particular, patients undergoing this

procedure should be started on aspirin before the procedure or be bridged with heparin. A transesophageal echocardiogram (TEE) is typically done simultaneously with the cardiac catheterization to ensure good positioning of the device and no residual defect or obstruction of other valves. Femoral venous percutaneous access is preferred. Arterial access is optional in these cases.

Initially, associated abnormalities including pulmonary venous abnormalities or associated atrial septal defects should be evaluated by TEE. Pressure and flow measurements are first obtained using a Berman or multiuse catheter. The defect is then crossed by a catheter and a wire is positioned in the left upper pulmonary vein through the defect to serve as an anchor. As the septal defects often stretch and are not symmetrical, balloon sizing is regularly performed. A balloon stretched and sized ASD is usually 30% larger than the measured TEE dimension prior to intervention. Balloon sizing also allows for temporary occlusion of the
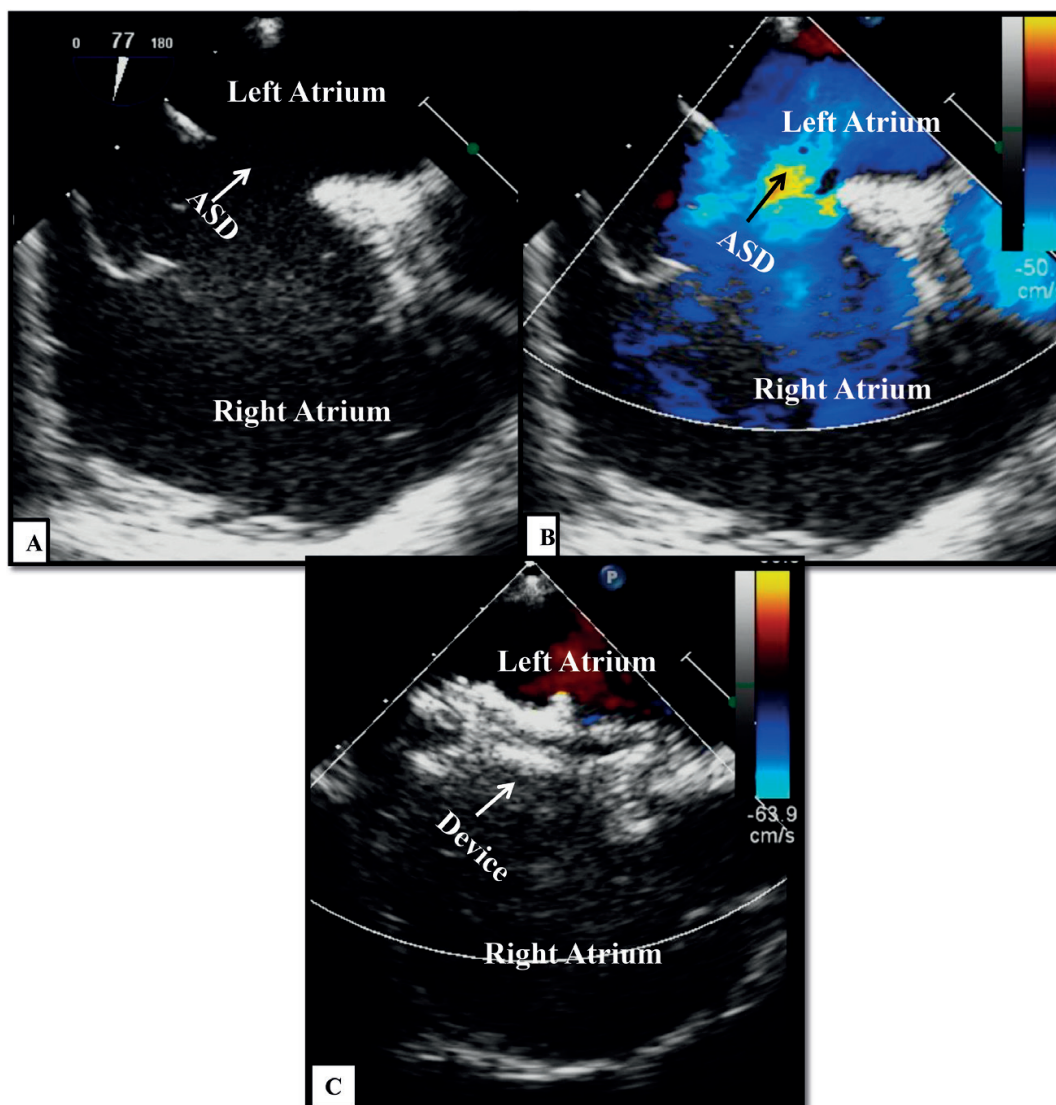


**Figure 7.** Device closure of an ASD in a 10 y/o asymptomatic patient found to have moderate right atrial and right ventricular dilation. (A) The right atrium is moderately dilated due to a large ostium secundum atrial defect. (B) Color flow demonstrated all left to right shunt (blue color flow in the image). (C) Color Doppler after deployment of a two disk Amplatzer septal occluder shows no residual left to right shunting and complete closure of the ASD.
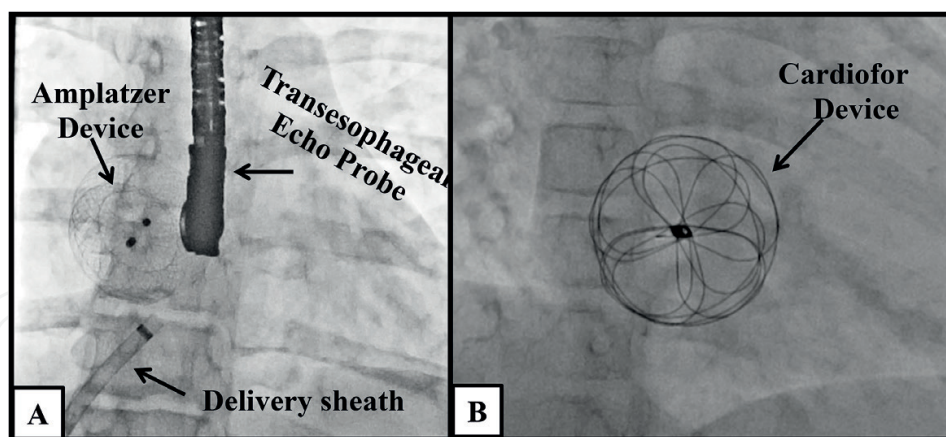
**Figure 8.** Fluoroscopic images of different deployed ASD closure devices. (A) Amplatzer septal occluder post deployment along with its delivery sheath. ASD closures are typically done under transesophageal echocardiographic guidance. (B) Cardioform septal occluder.

ASD to determine its hemodynamic effects on the heart and whether the patient will tolerate closure. Once this is done, a device is advanced through a sheath to the defect. Most popular devices consist of a two disk system [Gore Helex Septal Occluder (HSO) (W.L. Gore, Flagstaff, AZ) and Amplatzer septal occluder (ASO) (AGA Medical, Golden Valley, MN)]. The distal disk is deployed in the left atrium and pulled back until it is snug against the atrial septum. The proximal disk is then deployed and the device is released (**Figures 7** and **8**). Post procedure repeat chest films and echocardiograms over the next 24 hours are important to verify secure device positioning [12].

### 5.4. Complications

Reported severe complications in one study was as high as 7%, which is less than that of surgical closure [13]. During deployment of an occlusion device, components rotate and then return to their inherent shape. Device embolization is an unlikely complication but serious complication that occurs in the first 24–48 hours of the procedure. Erosion of a device into the adjacent anterosuperior wall or aorta is 0.1% in the US [14]. Incidence of device-associated thrombus is 1.2% and primarily occurs within the first month of the procedure. For this reason, patients are typically placed on a regimen of aspirin and clopidogrel for 3–6 months. Arrhythmias, primarily atrial fibrillation can occur with any manipulation and scarring of the atrium. The post-procedure incidence of atrial fibrillation is 6% [15]. Additionally, endocarditis prophylaxis is recommended for 6 months after device closure [12].

## 6. Balloon atrial septostomy

### 6.1. Introduction and indication

Balloon atrial septostomy, otherwise known as the Rashkind intervention, was described nearly 50 years ago as a life-saving emergent procedure required in particular congenital heart disease. Rashkind first described a transvenous approach for an atrial septostomy

in patients with transposition of the great arteries in the early 1960s. The procedure itself involves either widening an existing restrictive atrial level communication (**Figure 9**), or perforating the septum to manufacture a communication. Although the procedure is emergent, it can be planned even before birth based on fetal echocardiography. The primary purpose of an atrial septostomy is to enhance atrial mixing, decompressing the left atrium in a left sided obstructive lesion, or augmenting cardiac output in a right sided obstructed lesion. Primary congenital heart diseases that may require a balloon atrial septostomy include single ventricle physiology such as hypoplastic left heart syndrome and tricuspid atresia, transposition of the great arteries without a ventricular septal defect, and total anomalous pulmonary venous return.

### 6.2. Catheter procedure

The procedure can be performed in either the intensive care unit or a cardiac catheterization laboratory, the latter being preferred. In some centers, the infants are delivered in the cardiac catheterization lab to avoid any desaturation and cerebral hypoxia from delay. Access may someones be possible through the neonatal umbilical vein. If there are venous duct tortuosities, access through a femoral venous approach is required. Transthoracic echocardiographic guidance is highly recommended. Newborns are too small for a transesophageal echocardiogram. The patient should be anticoagulated with heparin and receive antibiotic prophylaxis at birth.

The procedure begins with advancing a balloon catheter into the right atrium and into the left atrium through the restrictive septum. In some cases, congenital heart disease such as hypoplastic left heart syndrome can have a thick atrial septum without any communication. In this case, a transseptal puncture needle or radiofrequency wire perforation may be required. Once the balloon is in the left atrium, it is inflated and, with a quick short movement, is passed back to the right atrium where it is deflated (**Figure 10**). This procedure is repeated several times and the presence of an atrial shunt is confirmed by echocardiography (**Figure 11**). In rare cases, a stent can be placed in the interatrial communication for a long-lasting result [16].
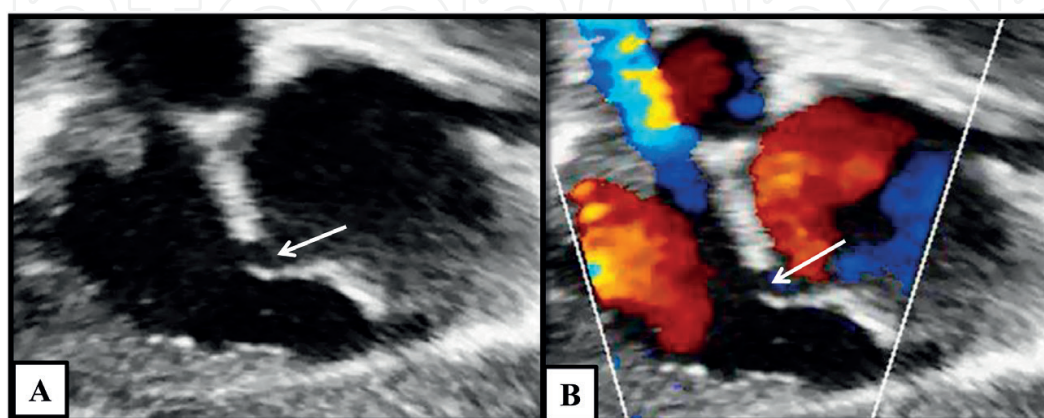


**Figure 9.** Transthoracic echocardiogram of a term newborn infant with D-transposition of the great vessels and a restrictive atrial septum. (A) 2D image of the restrictive atrial septal defect with bowing of the atrial septum. (B) There is nearly absent color Doppler signal across the defect representing scarce blood flow across the defect.
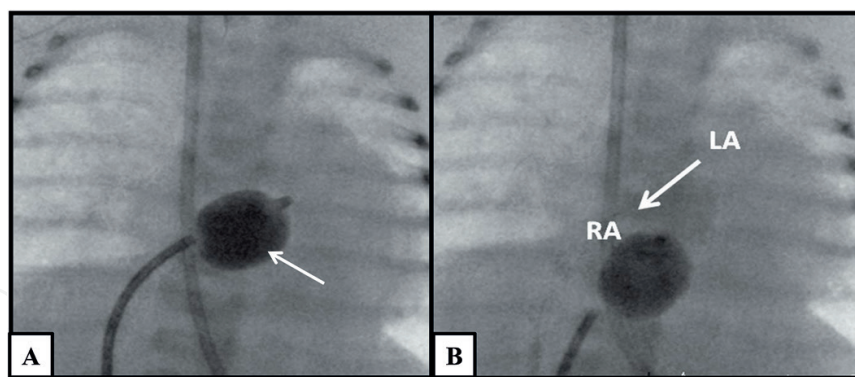
**Figure 10.** Fluoroscopy of a balloon atrial septostomy in the same patient. (A) The balloon is inflated while positioned in the left atrium with the catheter through the restrictive atrial septal defect. (B) In a swift motion, the inflated balloon is pulled back to the right atrium to dilate the atrial septal defect. This process is repeated a number of times.
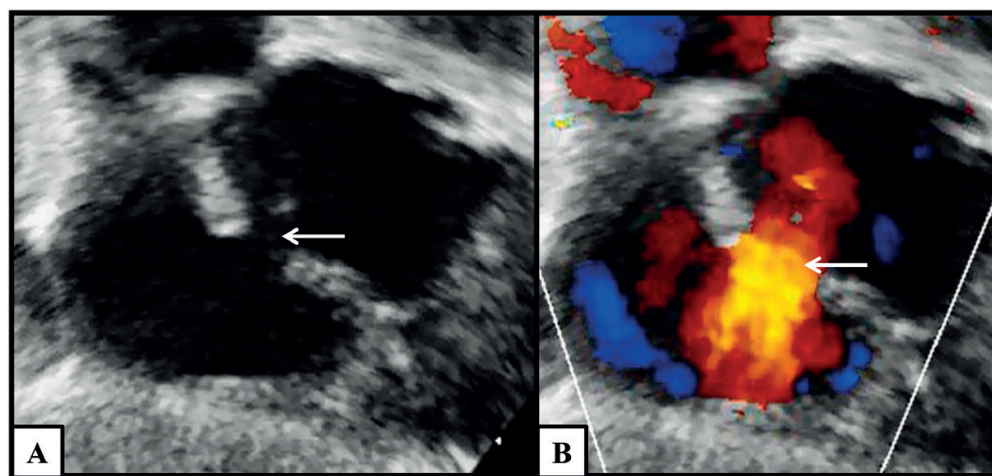


**Figure 11.** Post atrial septostomy echocardiogram of the same patient. (A) The atrial septal defect has now been dilated. (B) Color Doppler reveals unobstructed left to right shunting across the newly widened defect.

### 6.3. Complications

Balloon atrial septostomy is a life-saving emergent procedure with rare complications. Rupture of the balloon with embolization of the fragments can occur and require retrieval [17]. Traumatic complications including damage or rupture of the atrial appendage, mitral valve, or pulmonary veins can occur. Stroke has been reported, but recent studies have shown no increase odds of brain injury in patients who undergo a balloon atrial septostomy [18]. The most common complications include transient cardiac arrhythmias that require rapid treatment but generally resolve [16].

## 7. Coarctation of the aorta

### 7.1. Introduction

Coarctation of the aorta involves narrowing of various segments of the aorta. This often results in pathologic obstruction of systemic blood flow. Coarctation is the fourth most common

congenital heart defect. It comprises of 7% of all congenital heart disease and affects ~0.04% of live births. Males are twice as likely to have coarctation compared to girls [19]. The more severe the narrowing, the more symptomatic a child will be, and the earlier the problem will be noticed. In some cases, coarctation is noted in infancy. In others, however, it may not be noted until school-age or adolescence. Symptoms may include diminished blood pressures in the lower extremities, diminished pulses in the lower extremities, and signs of left sided heart failure including poor feeding, poor weight gain, and cool extremities. In severe cases of coarctation of the aorta, babies may present with hypoxia [20].

Initially, transcatheter balloon angioplasty was introduced in the 1980s as an adjunctive treatment to recurrent coarctation of the aorta following surgical therapy. Both treatments had flaws including re-coarctation of the aorta, residual hypertension, dissection of the aorta, and other aortic wall damage including aneurysm formation [21, 22]. To reduce complications and limitations, balloon-expandable stent therapy was introduced as a novel treatment for congenital heart diseases including coarctation of the aorta in the early 1990s [23]. Since that time, stent therapy has become widely accepted in children and adults for native and re-coarctation of the aorta [24].

## 7.2. Indications

The primary indication for repair in an older patient is an upper to lower-extremity systolic gradient of greater than 20 mmHg and/or the presence of symptoms including claudication and headaches. Of note, accurate blood pressure measurements are of the utmost importance. Ideally, three consecutive measurements in each extremity is required. Additional criteria include a diameter to diameter ratio of less than 0.6 between the narrowest portion of the coarctation of the abdominal aorta [25]. Cardiac balloon angioplasty and stent placement is rarely performed in small infants presenting with coarctation as rapid growth of the infant results in function coarctation from small stents.

## 7.3. Catheterization procedure

All patients should have standard catheterization precautions with general anesthesia, antibiotic prophylaxis, and anti-coagulation with heparin. The procedure occurs with a retrograde transfemoral approach by accessing both the femoral artery and vein. Care is taken to access the artery as proximal as possible and to avoid access past the bifurcation of the femoral artery. This ensures access to a large diameter vessel as larger stents require larger sheaths that can increase the risk of vascular compromise of the respective limb.

All patients undergo a complete right and left-heart catheterization as well as a detailed angiogram of the aorta using one of many various angiographic catheters such as a pigtail or multitrack catheter. Rotational angiography can provide additional imaging details of the aorta. Evaluating each branching vessel and its relationship to the coarctation segment is essential. Typically, a stiffer guidewire is positioned across the area of stenosis in preparation for stent placement.

Experience of the interventional cardiologist is of the utmost importance when choosing a stent. Typically, the diameter of the stent should not exceed the abdominal aorta diameter. Additionally, the ratio of the stent diameter to the narrowest coarctation segment should

be less than 3.5 [26]. Once the stent is chosen, it is loaded over a balloon-in-balloon catheter. The inner balloon allows repositioning after partial deployment of the stent. Successful treatment is defined as a residual gradient less than 10 mmHg and post intervention vessel caliber of greater than 80% of the normal adjacent aortic segment (**Figure 12**). Per the revised 2015 Bethesda guidelines, patients who have intravascular stent placement should avoid contact sports for 3–6 months post procedure. Patients often require continued anti-hypertensive medications. They require antiplatelet therapy and endocarditis prophylaxis for up to 6 months post procedure [26].

### 7.4. Complications and stent/balloon angioplasty versus surgery

Outcomes are generally excellent in the acute and long-term setting. Persistent hypertension is noted in up to 23% of patients requiring continued anti-hypertensive medication. Aortic wall complications are rare but can be severe and lead to cardiovascular compromise. These include dissection and rupture. In a cohort of 565 procedures, there was a 1.6% rate of aortic wall pathology especially in patients who underwent pre-stent angioplasty, balloon angioplasty ratio > 3.5, and age greater than 40 years [27].

Aortic aneurysm is a complication in 5–9%. Most are small and do not require procedural re-intervention, but those that are progressive or large can be treated with placement of a covered stent [27, 28]. Cerebrovascular accidents are exceedingly rare with a rate of less than 1% [29].

Various studies have compared outcomes between surgical repair, balloon angioplasty, and stent placement as treatments for coarctation of the aorta. A study by Forbes et al. compared



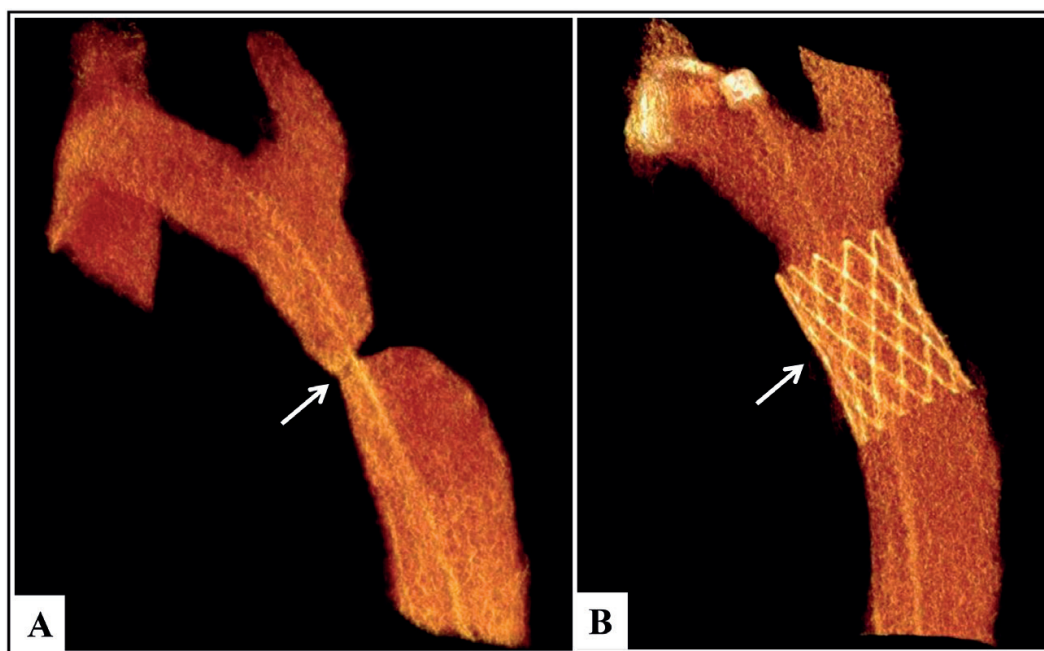**Figure 12.** Three-dimensional reconstruction of a rotational angiogram in an 8-year-old boy with severe coarctation of the aorta who underwent stent placement. (A) There is discrete stenosis at the aortic isthmus. A stent loaded balloon catheter can be seen across the narrow portion and into the ascending aorta. (B) Resolution of the angiographic coarctation with placement of a bare metal stent.

intravascular stent placement, balloon angioplasty, and surgical repair in a cohort of 350 patients and found that stent patients had fewer acute complications though at intermediation follow up there was no significant difference in persistent hypertension. There was a significantly higher ratio of the post intervention coarctation are to the adjacent descending aorta and a lower rate of aortic wall injury in the patients with intravascular stents compared to those with balloon angioplasty and surgical repair [25].

# 8. Aortic stenosis

## 8.1. Introduction

The aortic valve is a semilunar valve that connects the left ventricle to the aorta in the normal heart. The major difference between the aortic valve and pulmonary valve is the typical absence of an infundibular septum. Aortic stenosis occurs with obstruction below the valve (subvalvar), at the valve (valvar), and above the valve into the left ventricular outflow tract (supravalvar). Aortic stenosis accounts for 3–8% of all congenital heart disease. Valvar stenosis is the most common comprising of 75% of aortic stenosis. Of note, this is typically associated with either a bicuspid valve where two of the 3 cusps are fused, or a unicuspid valve where all three cusps are fused. Aortic stenosis is also associated with various other congenital heart defects as well as genetic syndrome such as Williams syndrome. Symptoms of aortic stenosis are similar to those of any left ventricular outflow tract obstruction including chest pain or syncope with exercise and dyspnea on exertion. There may also be respiratory distress in infants caused by pulmonary edema, though this is typically absent in children and adults [30].

As with most other valvular disease, diagnosis is primarily made by an echocardiogram which is ordered based on clinical suspicion. Echocardiogram can demonstrate the morphology of the valve and characterize the location and degree of obstruction using both continuous wave and pulse wave Doppler, and color Doppler. Untreated aortic stenosis carries with it a 4% risk of endocarditis and a 5% risk of sudden death.

## 8.2. Indications

Aortic stenosis with less than a transvalvar gradient of 50 mmHg is typically well tolerated. Aortic stenosis is a progressive disease with between 21 and 41% of these patients progressing to severe aortic stenosis defined as a gradient of greater than 50 mmHg. There is an associated 1% per patient-year risk of sudden death in these patients. Balloon aortic valvuloplasty is typically the first line treatment for patients with valvar aortic stenosis with a pressure gradient of greater than 50 mmHg and minimal aortic regurgitation. In patients with subvalvar and supravalvar stenosis, balloon angioplasty is typically not effective. Surgical repair by resection of the areas of stenosis is primarily the treatment for subvalvar and supravalvar stenosis.

## 8.3. Catheterization procedure

Most patients should have standard catheterization precautions with general anesthesia, antibiotic prophylaxis, and anti-coagulation with heparin. In some cases outside of the neonatal

period, the procedure can be performed under conscious sedation. The procedure occurs with a retrograde transfemoral approach by accessing both the femoral artery and vein. Care is taken to access the artery as proximal as possible and to avoid access past the bifurcation of the femoral artery.

This ensures access to a large diameter vessel as larger sheaths that can increase the risk of vascular compromise of the respective limb.

All patients undergo a complete right and left-heart catheterization as well as a detailed angiogram of the aorta and aortic valve. The most important measurement in a transvalvar gradient that can be obtained by a pullback measurement with a catheter, or simultaneous measurements in the aorta and left ventricle using either a trans-atrial-septal approach (in the presence of an ASD or PFO) or an additional arterial access point. The balloon size chosen should be 80–90% of the annulus size to reduce the risk of significant aortic regurgitation.
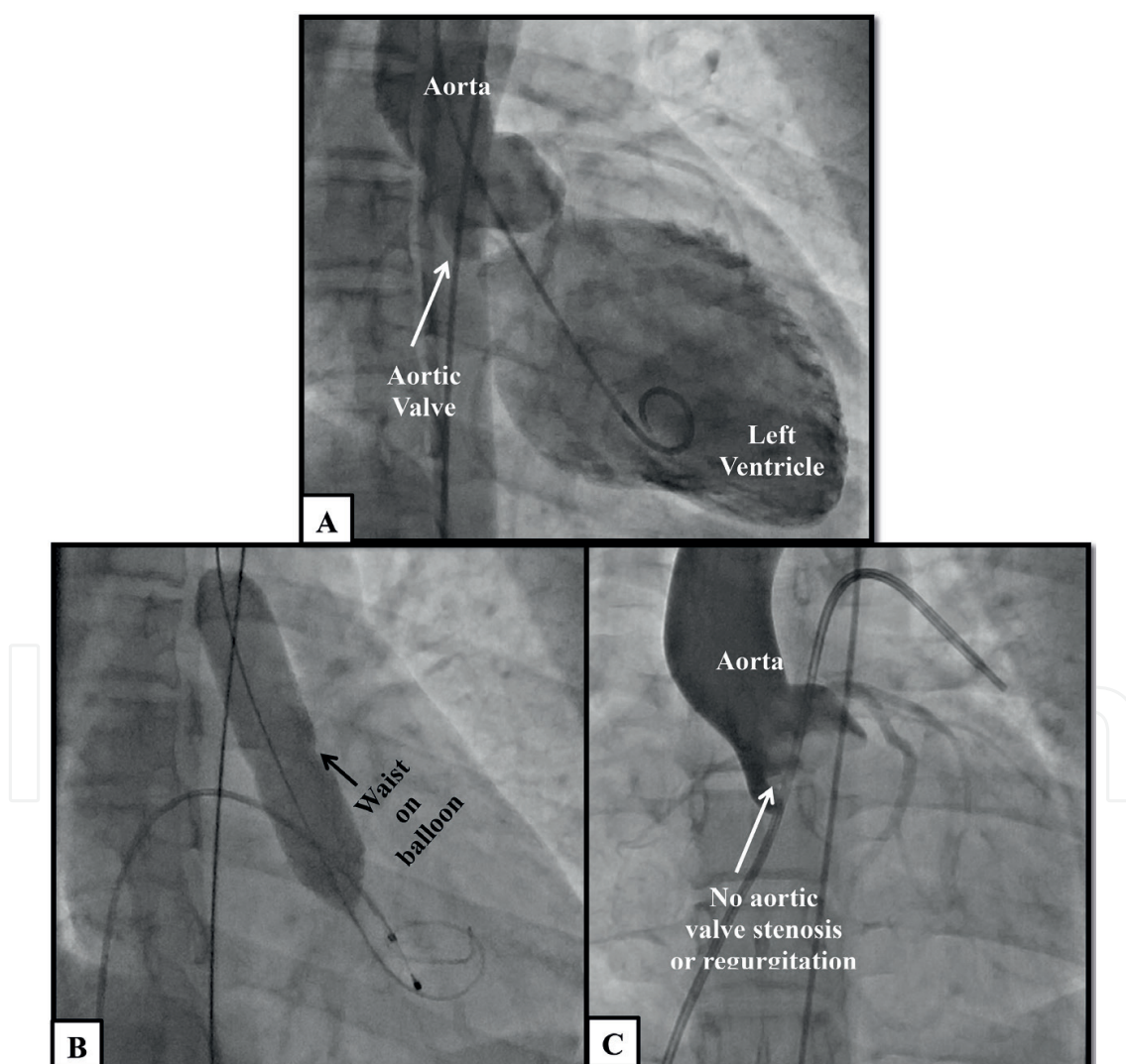


**Figure 13.** Aortic balloon valvuloplasty in a 14 year old patient with moderate aortic valve stenosis and dizziness. (A) There is moderate stenosis at the level of the aortic valve. A pigtail catheter rests in the left ventricle retrograde from the aorta. (B) During balloon angioplasty of the valve, a "waist" is seen at the area of stenosis. (C) Post valvuloplasty there is no evidence of residual stenosis or aortic regurgitation.

Once the procedure is complete, the gradient is remeasured. Typically, a successful angioplasty results in reducing the gradient by half without a significant increase in aortic regurgitation (**Figure 13**).

### 8.4. Complications

The most common complications with balloon aortic valvuloplasty are aortic regurgitation and injury to the peripherally accessed vessel. Significant aortic regurgitation occurs in approximately 10% of all aortic valve balloon dilations which is comparable to rates in surgical valvotomy. Oversizing of the aortic balloon to be greater than the size of the aortic valve annulus is a primary risk factor. In patients with balloon ratios greater than 1.0 (I.E. a balloon larger than the diameter of the aortic valve annulus), at least moderate aortic regurgitation is seen in 25% of patients. Vascular injury is less common with improvement in techniques and availability of monitoring equipment [31]. Re-intervention is common and is reportedly required in up to 50% of patients. Typically, each subsequent intervention carries a higher risk of complications. The incidence of moderate aortic regurgitation in repeat dilations is at least 25% [32].

## 9. Pulmonary stenosis

### 9.1. Introduction

The pulmonary valve is a semilunar valve that connects the right ventricle to the pulmonary artery in a normal heart. Pulmonary stenosis is primarily a sequelae of congenital heart disease but can be acquired due to surgical or transcatheter interventions. Stenosis can occur at the level of pulmonary valve, above the pulmonary valve, or below the pulmonary valve. Valvular stenosis is the most common, accounting for up to 90% of cases [33]. It is often an isolated finding, but can be associated with congenital heart lesions, most notably tetralogy of Fallot. It is also associated with genetic conditions such as DiGeorge syndrome and 22q11 deletion, Williams syndrome, Alagille syndrome, congenital rubella, and Noonan's syndrome. Noonan syndrome is particularly important as it results in dysplastic, thickened pulmonary valve leaflets that typically do not respond well to catheter-based balloon valvuloplasty. Patients with tetralogy of Fallot are more likely to have a component of sub-valvar and infundibular stenosis [34].

Patients with mild isolated pulmonary stenosis are typically asymptomatic and the stenosis rarely progresses rapidly. Symptoms are associated with the resulting decrease in cardiac output from stenosis. These include dyspnea on exertion, fatigue, and right ventricular hypertrophy, right ventricular diastolic and systolic dysfunction. Patients with severe pulmonary stenosis in the neonatal or early childhood can present with cyanosis in the setting of a intracardiac shunt such as an atrial septal defect.

Diagnoses by echocardiogram is the gold standard. Two-dimensional and three-dimensional echocardiography can evaluate the morphology of the stenosis and pulmonary valve, and evaluate the function and size of the right ventricle. Continuous and pulse wave Doppler can

help determine pressure gradients and color Doppler can help quantify and resultant pulmonary regurgitation. In some cases, MRI can accurately assess the degree of regurgitation, flow across the valve, and measure right ventricular size [35].

The three main treatment modalities for pulmonary valve stenosis are balloon pulmonary valvuloplasty, surgical valvotomy, and pulmonary valve replacement, either by surgery or cardiac catheterization. The first balloon valvuloplasty of the pulmonary valve was done in 1979 and is by far the procedure of choice. Valvuloplasty is generally safe and effective in greater than 90% of patients [36].

## 9.2. Indications

Balloon pulmonary valvuloplasty is indicated for asymptomatic patients with a domed valve morphology, a peak gradient by echocardiography of greater than 60 mmHg, and up to moderate pulmonary regurgitation. Symptomatic patients are treated with an echocardiographic gradient of greater than 50 mmHg and less than moderate pulmonary regurgitation. Immediate effects of balloon valvuloplasty include improved longitudinal right ventricular motion, reduced dyssynchrony in pediatric patients, reduction of tricuspid regurgitation, and improvement in the pulmonary valve gradient. Patients with supravalvar and subvalvar pulmonary stenosis may undergo balloon valvuloplasty but is often less effective, primarily in patients with Noonan syndrome.

## 9.3. Catheterization procedure

All patients should have standard catheterization precautions with general anesthesia, antibiotic prophylaxis, and anti-coagulation with heparin. Percutaneous femoral venous access is the preferred entry site for balloon pulmonary valvuloplasty. The procedure begins with pressure measurements of the right ventricular and pulmonary artery using a Berman or other multipurpose catheter. Pulmonary valve are can be calculated using the Gorlin formula. Simultaneous recording of the right ventricle and femoral artery pressure is done to assess severity of the obstruction. If the right ventricular systolic pressure is greater than 75% of the systemic systolic pressure, there is significant pulmonary valve obstruction. Angiography is an important tool in confirming the site of stenosis in addition to obtaining accurate measurements of the pulmonary valve annulus [37].

A guidewire is positioned in the distal left pulmonary or right pulmonary artery. In infants, the wire may be positioned in the descending aorta through the patent ductus arteriosus. The purpose of the wire is to guide the balloon catheter and increase its stability. A number of different balloon catheters are available and are often chosen based on availability and experience of the interventional cardiologist. The diameter of the balloon should be 1.2–1.4 times the size of the pulmonary valve annulus with a recommended ratio of 1.2–1.25 [38]. In cases of a dysplastic pulmonary valve, as is typically found in Noonan's syndrome, a ratio of 1.4–1.5 may be used [39]. Successful balloon valvuloplasty is defined as a residual gradient of less than 30 mmHg. Patients with successful valvuloplasty may return to competitive sports 3–6 months post procedure (**Figures 14** and **15**).
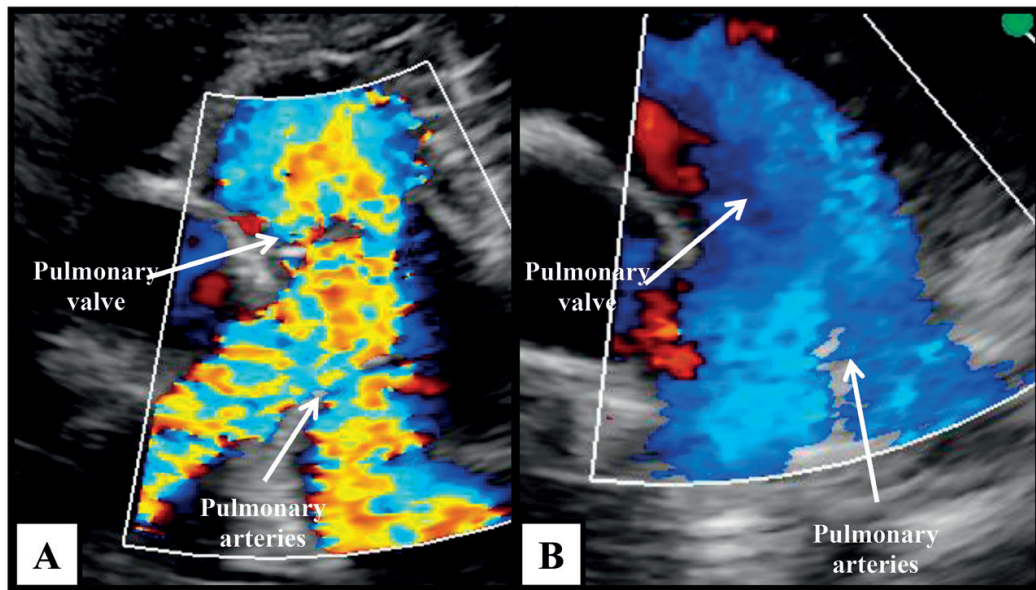
**Figure 14.** Color Doppler transthoracic echocardiogram of a 2 year old patient with trisomy 21 and pulmonary stenosis who underwent balloon pulmonic valvuloplasty. (A) There is flow acceleration (blue and red color speckling) across the pulmonary valve indicating valvar stenosis. (B) Post balloon pulmonic valvuloplasty there is no flow acceleration or signs of obstruction.
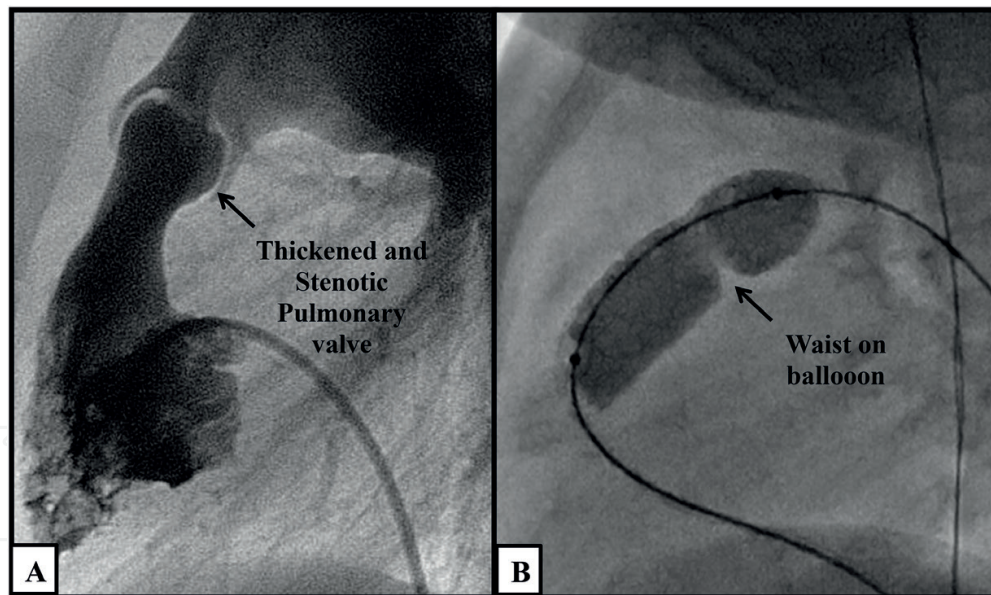


**Figure 15.** Fluoroscopic images of the above patient during a balloon pulmonic valvuloplasty. (A) There is a thickened and stenotic pulmonary valve typical of patients with Noonan syndrome. (B) During balloon valvuloplasty a "waist" at the area of stenosis is seen.

### 9.4. Complications

Severe complications are rare occurring in 0.6% of cases and mortality in 0.2% [40]. A major complication is transient but severe right ventricular outflow tract obstruction (sometimes

known as a "suicide right ventricle") that can occur immediately after valvuloplasty. Long standing or severe pulmonary stenosis results in right ventricular hypertrophy. When the stenosis is resolved and the systolic right ventricular pressure drops, the hypertrophy results in acute narrowing of the outflow chamber and must be treated with fluid resuscitation and beta blockade [41].

The most common long term side effect is pulmonary regurgitation and recurrent pulmonary stenosis [42]. One study found that 60% of patients followed for 10 years after pulmonary valvuloplasty had at least moderate pulmonary regurgitation. However, pulmonary regurgitation was well tolerated in the majority of patients. Only 5% had right ventricular dysfunction and 3% required intervention [43]. The lower the balloon:annulus ratio, the less risk of significant pulmonary regurgitation. The Long term impact of chronic pulmonary regurgitation from pulmonary valvuloplasty is not well studied. Surrogate data from pulmonary regurgitation secondary to surgical valvotomy or valvectomy suggests that right ventricular function begins to deteriorate after the first several decades [44].

Restenosis occurs in up to 25% of patients who undergo valvuloplasty defined as a peak gradient of >36 mmHg in 1–2 years post procedure. Around 15% of patients who have restenosis will require re-intervention with either surgery or repeat balloon pulmonary valvuloplasty. Risk factors for recurrent stenosis include a dysplastic valve, small annulus, high initial gradient, and a small balloon:annulus ratio defined as less than 1.2 [45].

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