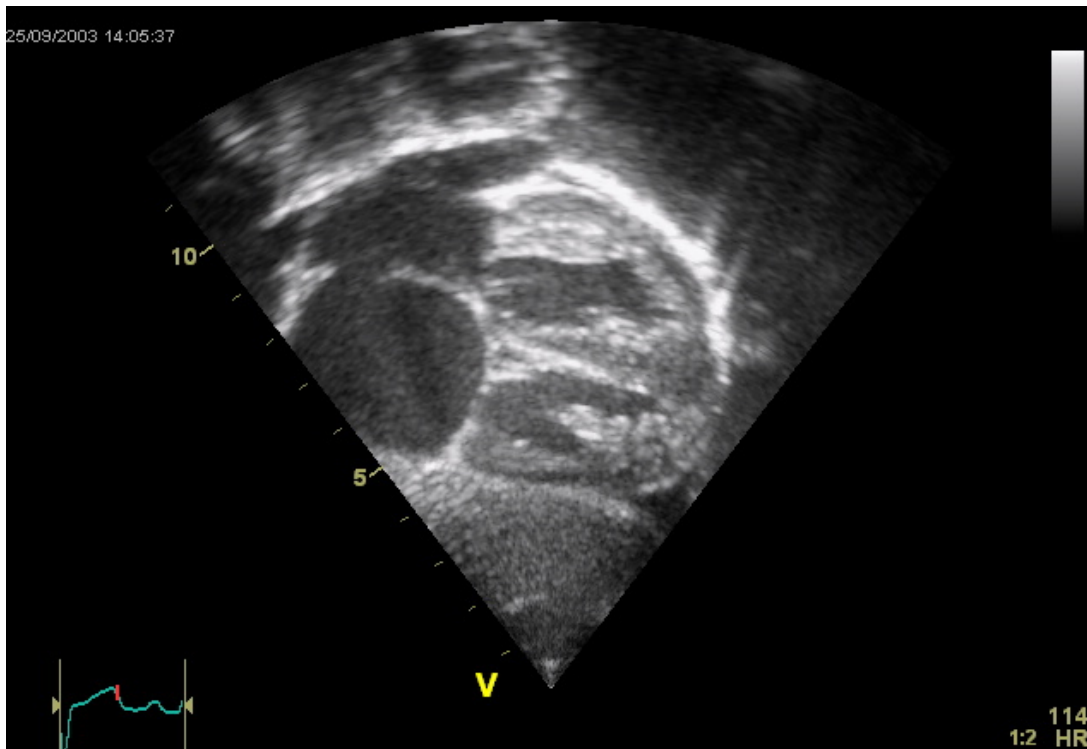


Pediatric Atrial Septal Defects — Classification and Surgery

[See online here](#)

Atrial septal defect and patent foramen ovale, represent diverse congenital heart diseases with the common denominator being a defect of closure of atrial septum. Embryologic development of atrial septum is essential in order to understand anatomic, pathophysiologic and clinical presentation of ASD that presents as asymptomatic, usually in children, to symptomatic forms that are mainly found in adults with long-term complications such as atrial arrhythmias, pulmonary hypertension, and heart failure. Clinically symptomatic patients present with fatigue, dyspnea, exercise intolerance, and atrial arrhythmias. Management options, when needed, include defect closure either by transcatheter device closure or open surgery procedure.

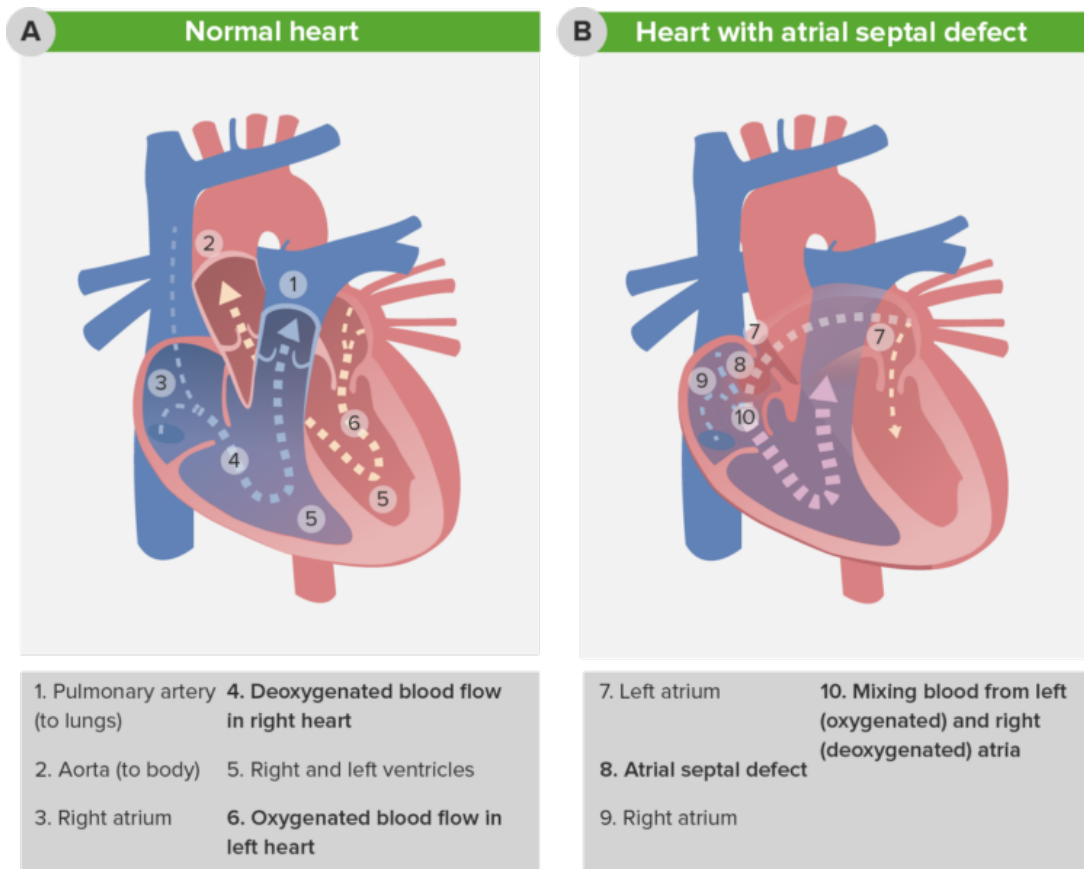


Definition of Pediatric Atrial Septal Defects

Atrial septal defect (ASD) is a **congenital heart condition** presented as a defect in the atrial septum that accounts for up to 10 % of all coronary diseases and up to 30 % of congenital defects recognized in adults. There is also another congenital heart disease named **patent foramen ovale** found in 25 to 30 % of the population that involves defects inside the atrial septum in the position of the foramen ovale.

High-Yield: Atrial septal defects encompass a group of conditions that are characterized

by the presence of an opening between the two atria. This opening can be by the patency of a normal opening, a patent foramen ovale, or by the presence of communication within the septum between the two atria.



By Lecturio.

Epidemiology of Pediatric Atrial Septal Defects

Atrial septal defects have an **estimated incidence of between 0.67 to 2.1 per 1000 live births**, which makes them the second most common form of congenital heart disease. The most common form of atrial septal defects is a **secundum atrial septal defect** which is identified in up to 90 % of cases. Sinus venosus and primum atrial septal defects are equally responsible for the remainder of the cases.

Atrial septal defects are associated with low mortality and morbidity. The mortality rate of atrial septal defects is estimated to be around 1 %.

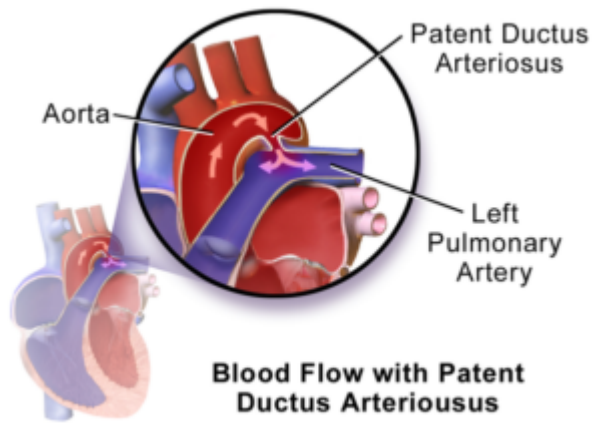


Image: "Patent Ductus Arteriosus." . By Blausen.com staff (2014).
"Medical gallery of Blausen Medical 2014". Wikijournal of
Medicine 1 (2). DOI:10.15347/wjm/2014.010. ISSN 2002-4436..
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Infants with large atrial septal defects but no patent ductus arteriosus or ventricular septal defects usually are symptomatic and can fail to thrive. Atrial septal defects have also been associated with an **increased risk of atrial arrhythmias such as atrial fibrillation**. Atrial septal defects are twice more common in girls than in boys.

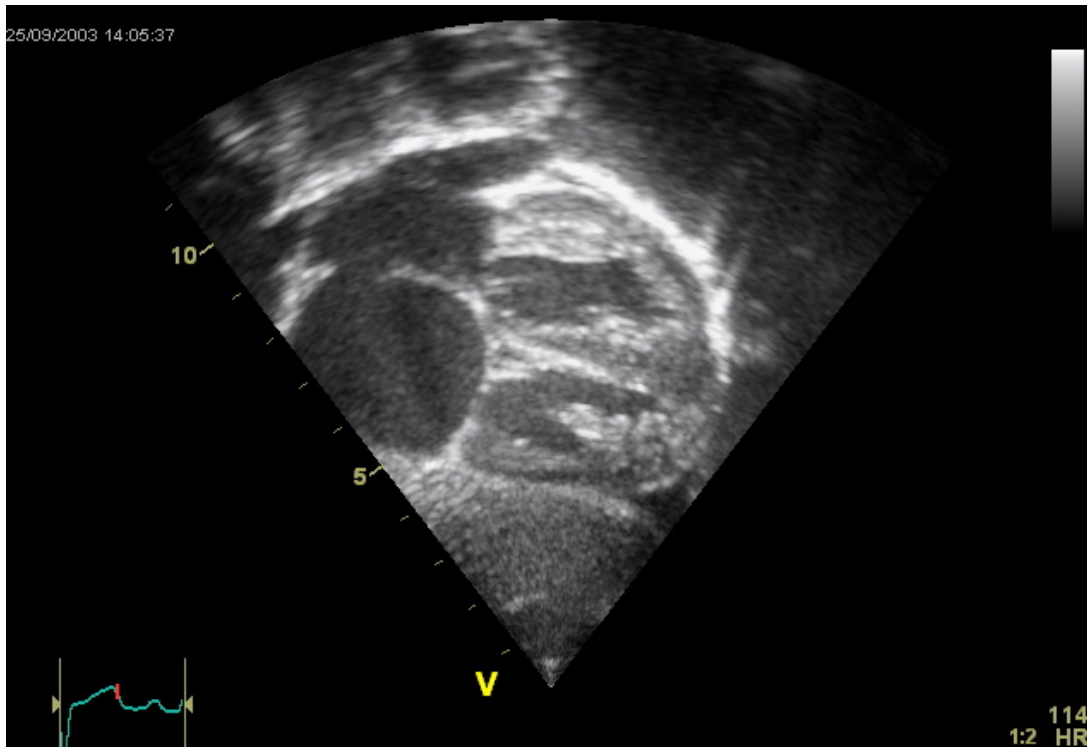
The most common age of diagnosis of atrial septal defects is during adulthood. Patients with large atrial septal defects might have an audible murmur on heart auscultation when they are a couple of months old but rarely at birth.

Classification of Pediatric Atrial Septal Defects

Four main types of atrial septal defects have been described:

1. Ostium Secundum Defect

The ostium secundum defect is the most common type of ASD, accounting for more than 75 % and up to 90 % of all cases, with defect positioned in fossa ovalis (middle part of interatrial septum). Usually, ASD is in form of a single defect but there have been reported cases of fenestrated defects. Associated anomalies may include mitral valve prolapse and anomalous pulmonary venous return.



ECG of an ASD (Ostium Secundum)

2. Ostium Primum Defect (*known as endocardial cushion defect*)

Ostium Primum defect is a less common type of ASD which comprises of about 15 % of cases. The defect is positioned in the lower part of the interatrial septum (the level of endocardial cushion). The defect is often associated with a defect in the basal part of the ventricular septum which leads to abnormalities of the mitral valve and the tricuspid valve such as the single atrioventricular valve as well as the cleft mitral valve. An important associated extra-cardial pathology is Down's syndrome.

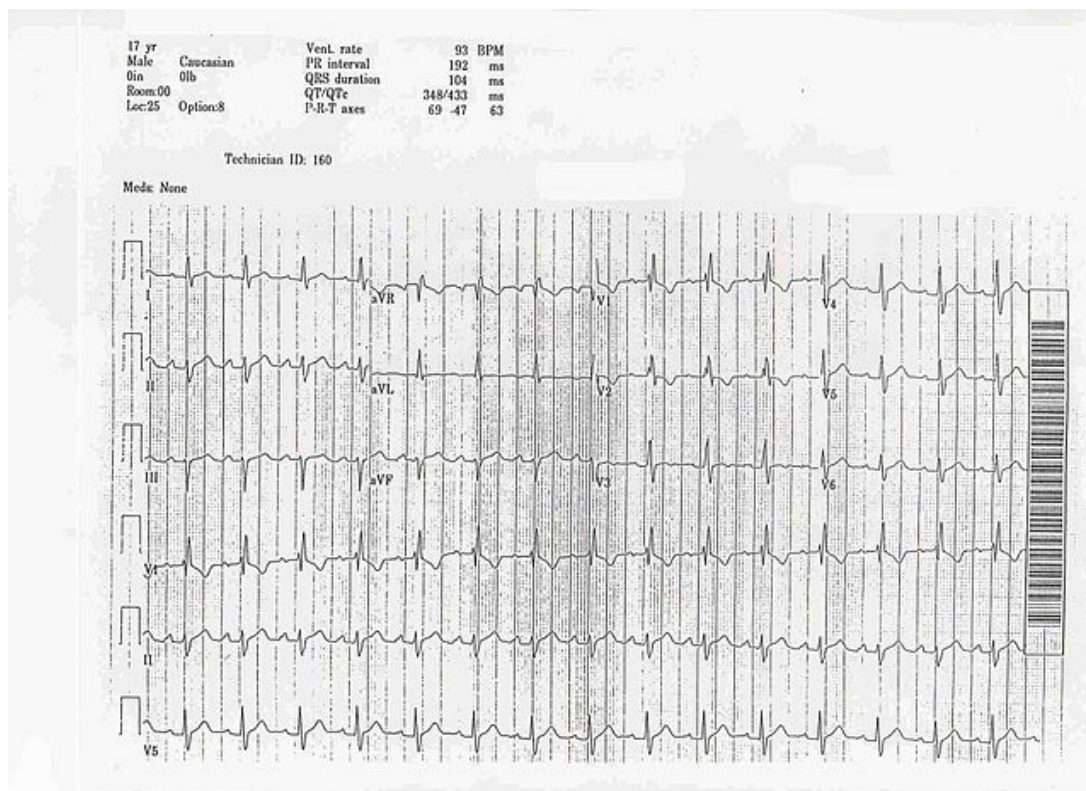


Image: "ECG of a patient with Ostium primum ASD". By WikiSysop - http://askdrwiki.com/mediawiki/index.php?title=ASD_Primum_Defect. License: [CC BY-SA 3.0 SA](https://creativecommons.org/licenses/by-sa/3.0/)

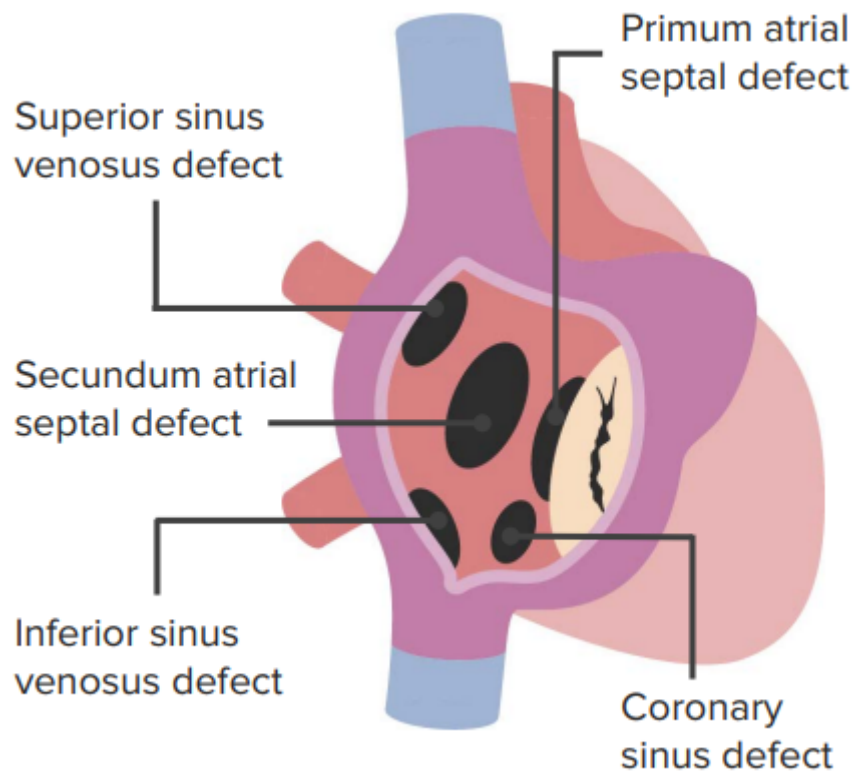
3. Sinus Venous Defect

Sinus Venous defects account for up to 10 % of all ASD cases and are presented by a defect in the upper atrial septum that involves the superior vena cava vein junction, or in the rare cases inferior vena cava vein. The most common manifestation of sinus venous defect is anomalous pulmonary venous return, mainly the right upper pulmonary vein.

4. Coronary Sinus Defect

Coronary Sinus defect is a rare form of interatrial communication presented as a variable size defect between the coronary sinus and the left atrium. It is in majority of cases associated with a persistent left superior vena cava and an atrial septal defect of the coronary sinus type. This type of defect is in most cases part of a more complex cardiac malformation.

Most of the symptoms of atrial septal defects are attributed to left-to-right shunting. This can only happen with moderately large to large atrial septal defects but very rarely with small atrial septal defects.



By Lecturio

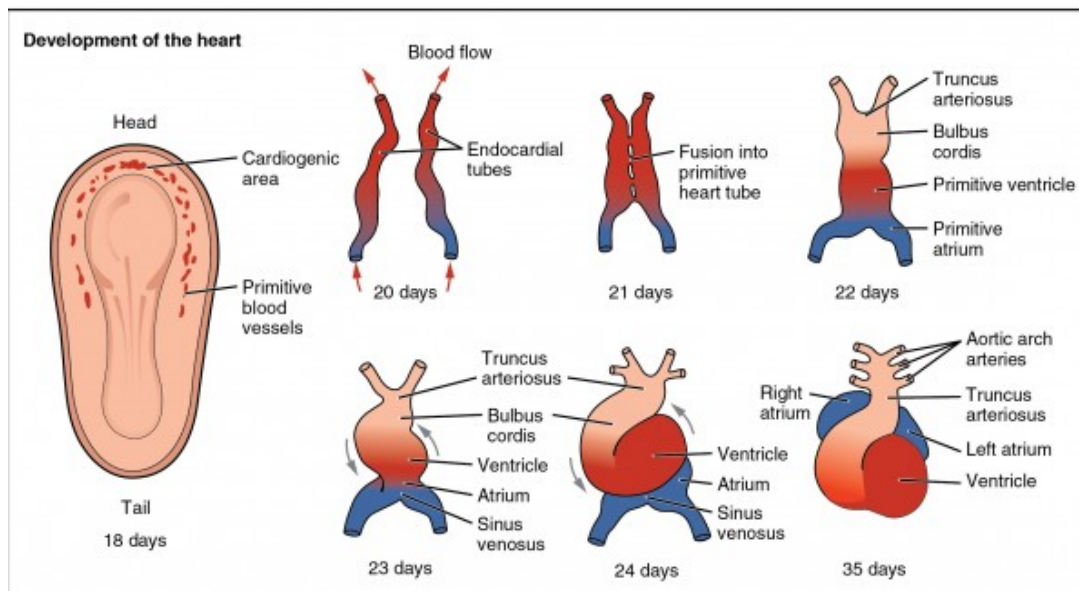
Pathophysiology of Pediatric Atrial Septal Defects

ASD is presented **twice as often in females** compared to males, most ASDs result from a spontaneous genetic mutation, but hereditary forms have been reported.

Understanding the basic knowledge of heart anatomy and embryology is important to understand the pathophysiology of ASDs.

Embryology

The development of the heart begins in the 18–19th day after fertilization.



Picture: "Development of the Human Heart" by Phil Schatz. License: [CC BY 4.0](https://creativecommons.org/licenses/by/4.0/)

The development of the septum primum begins from a posteriorly located crescent-shaped piece of tissue growing to become the initial barrier between the right and left atria. **Because of its shape, the septum primum does not completely occlude the area between left and right atria leaving the opening named the ostium primum (the first opening).** This, in fetal development, allows blood to be driven from the right atrium to the left.

The growth of the septum primum reduces the ostium primum. But prior to closure of the ostium primum the second opening named **ostium secundum (the second opening)** is formed in the septum primum. The ostium secundum also allows right to left shunt on the atrial level.

On the right side of the septum primum begins the formation of the septum secundum presented as a crescent shaped anteriorly orientated thick, muscular structure. During its growth, the septum secundum leaves an opening named the foramen ovale, which is continuous with the ostium secundum, without change in blood shunting direction.

As the size of ostium secundum grows, the septum primum closes and in the end presents a flap (named the valve of foramen ovale) covering the left side of the foramen ovale. It serves as a pressure-regulated valve between the left and right atria allowing one direction right to left shunt in cases of increased right atrium pressure.

Pulmonary circulation pressure is greater than systemic circulation pressure due to non-functioning fetal lungs meaning that the pressure inside the right atrium is higher than that of the left atrium, all leading to the opening of the valve of the foramen ovale. **The situation drastically changes at birth, following the reversal of interatrial pressure gradient thus leading to the closing of the foramen ovale's valve. This closure is, in the beginning, functional, and during infancy becomes an anatomic closure.**

Normal cardiac anatomy results in complete separation of oxygenated and deoxygenated blood pools, leading to maintenance of one-on-one volume relationship between the systemic and pulmonary circulation.

Blood arriving from the systemic circulation through vena cava veins is deoxygenated, and by passing through the right atrium and right ventricle it is pumped inside the

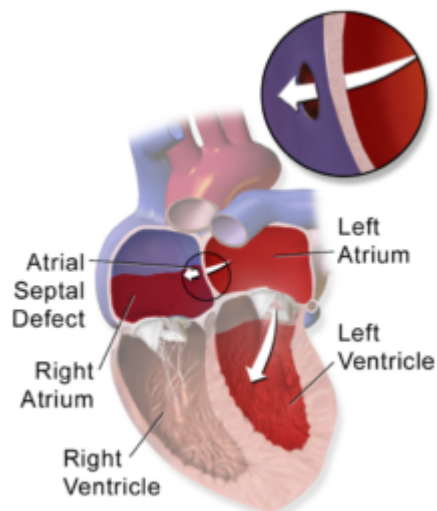
pulmonary artery to the lungs. Once oxygenated, the blood flows through the pulmonary veins to the left atrium, left ventricle and is pumped to the aorta then distributed throughout the entire organism, and by venous system, it is pumped into the right heart chamber.

A “shunt” represents an abnormal connection that allows direct blood flow from one side of the cardiac circulation to the other.

- A left-to-right (L-to-R) shunt means that the oxygenated blood is returned to the lungs without being pumped through the body.
- A right-to-left (R-to-L) shunt means the deoxygenated blood returns to the body without being oxygenated.

Both types of shunt lead to less efficient circulation and an overload of cardiac chambers. The severity of symptoms is mostly determined by the volume of shunted blood.

Pathophysiology



Heart with Atrial Septal Defect

Image: “Atrial Septal Defect.”. By Blausen.com staff (2014). “Medical gallery of Blausen Medical 2014”. Wikijournal of Medicine 1 (2). DOI:10.15347/wjm/2014.010. ISSN 2002-4436. License: [CC BY 3.0 SA](https://creativecommons.org/licenses/by/3.0/)

Focusing on ASD, it is observed that blood flow through the defect occurs in both systole and diastole. **Usually, a shunt is predominantly left to right, but a common scenario includes transient right-to-left shunts.** The main part of the shunt flow occurs during diastole, leaving blood in each atrium one of the two alternative pathways: either following the anatomical route through the ipsilateral AV valve direction or passing through the ASD and filling the opposite atrium and ventricle.

The shunt direction during diastole in ASD is dependent on differences in the compliance and capacity of the ventricles.

Ventricular compliance is multifactorial determined, but in the classical scenario, the left ventricle, because of working against systemic circulation high-pressure gradient, is thick walled and less compliant, thus leading to a preferential left to right direction of blood through the septal defect.

If an increase in the right ventricle afterload occurred, as in cases of either congenital obstructions in the pulmonary arteries/veins or in high pulmonary vascular resistance (pulmonary parenchymal disease or primary pulmonary hypertension), then the right ventricle becomes less compliant and the right to left shunt is present.

In the systole, AV valves are closed and ventricular compliance has no effect on blood flow direction across the defect, leaving flow volume and direction subject to, in the first place, the size of the defect (determinant in the volume) and atria's differential capacities or compliances (determinant of flow direction).

The presence of AV valve regurgitation affects systolic ASD flow direction, as well as significant mitral or tricuspid regurgitation.

In the end, **ASD's size determines the volume of shunting.**

Clinical Features of Pediatric Atrial Septal Defects

Although atrial septal defect is present at birth, **most babies will not have signs or symptoms of the disease.**

Most patients with atrial septal defects are asymptomatic during the first months and even years of life. Most patients are diagnosed incidentally by the identification of a heart murmur on a routine health maintenance examination.

Children with symptomatic atrial septal defects usually do not have any specific signs or symptoms. Exertional dyspnea, recurrent upper respiratory tract infections and failure to thrive are common symptoms seen with large atrial septal defects.

Children with a large atrial septal defect and a patent ductus arteriosus or who have an ongoing lung disease are more likely to have severe symptoms. This group of children might develop cardiac arrhythmias such as atrial fibrillation, embolic disease, or pulmonary artery hypertension. Congestive heart failure may also develop in this group of children.

A right ventricular cardiac impulse, palpable pulmonary artery pulsations, and a precordial bulge are common signs of atrial septal defects. When blood flow across the pulmonic valve is increased, a mid-systolic pulmonary ejection murmur may be heard. A fixed heart sound is commonly heard in children with atrial septal defects.

However, signs and symptoms of a significant or untreated atrial septal defect in children may present as follows:

- Tiring when feeding (infants)
- Difficulty breathing
- Shortness of breath when active or exercising
- Frequent respiratory or lung infections
- Skipped heartbeats or a sense of feeling the heartbeat
- A heart murmur, or a whooshing sound that can be heard with a stethoscope
- Swelling of legs, feet, or stomach area
- Stroke

In adult patients:

- Progressive shortness of breath with exertion
- Atrial arrhythmia due to stretching of the conduction system fibrillation
- Rarely a stroke/ systemic ischemic event because of paradoxical embolization

It is a common clinical scenario to diagnose atrial septal defect in an adult patient. Auscultation is a simple, yet effective technique for diagnosing it.

Diagnosis of Pediatric Atrial Septal Defects

Physical Examination

Auscultation may reveal an upper left sternal border mid-systolic murmur in combination with a wide and fixed splitting of the 2nd heart sound.

Patients with atrial septal defects usually have a normal complete blood count. Elevated levels of the brain natriuretic peptide have been linked to congestive heart failure and their levels should be checked in neonates presenting with heart failure and a large atrial septal defect.

Electrocardiogram (ECG)

ECG ranges from normal (in an uncomplicated ASD and small shunt) ECG to atrial arrhythmias (usually from the third decade) in form of atrial fibrillation, atrial flutter and supraventricular tachycardia.

The morphology of P waves is normal with secundum ASD's.

In sinus, venous ASD's P waves are associated with a leftward frontal plane P-wave axis (such as negative P in leads III and aVF and positive P in lead aVL), caused by an ectopic pacemaker due to near sinus node ASD's position.

The first degree AV block may occur in ASD irrelevant of the subtype, although it is most often present in ostium primum defects, accompanied by complete right bundle branch block and left anterior fascicular block, due to the ostium primum defect 's rim being in close relationship with the His bundle leading to abnormalities of impulse conduction through this area.

The QRS axis frontal plane is between $+95^{\circ}$ to $+135^{\circ}$ (presenting as right axis deviation). Left axis deviation frontal plane is sometimes encountered in uncomplicated secundum ASD, although it is a predictor of AV canal defect existence.

“Crochetage”: presents as a notch on the R wave in the inferior leads, is suggested to be a sensitive and specific ECG sign of ASD, correlating with both the size of the defect and degree of L-to-R shunt.

Chest X-ray



Image: "Abnormal chest X-ray as seen in a patient of Atrial septal defect." By WikiSysop. License: [CC BY-SA 3.0 SA](#)

Chest radiography may reveal a right atrial and ventricular dilatation, pulmonary artery dilatation and increased pulmonary vascular markings due to over circulation. Echocardiography with or without Doppler studies is essential in the diagnosis of atrial septal defects.

Classically, it can detect the presence of right atrial and ventricular dilatation, pulmonary arteries enlargement and also, if mitral regurgitation is present, left atrial enlargement may be seen; shunt vascularity is characterized by enlarged main pulmonary arteries and pulmonary vessels.

It is atypical in adult patients (usually over the age of 50) normal vasculature, pulmonary venous hypertension, left atrial enlargement, and even pulmonary edema may be present.

"Scimitar sign": the insertion of the pulmonary vein into the inferior vena cava in sinus venosus defect type of ASD presents as a right-sided paracardial density that is vertical and curved like a sabre.

Echocardiography

Echocardiography allows for the diagnosis of the defect and the evaluation of the degree of left to right shunting. It is preferable to obtain a subcostal oblique view of the heart in small infants and transesophageal echocardiography is recommended in older children and adults. Transesophageal echocardiography is also useful in excluding other cardiac abnormalities such as an unroofed coronary sinus or partial anomalous pulmonary return.

High-Yield: Echocardiography is the gold standard for the diagnosis of ASD.

- Transthoracic echocardiography defines diagnosis in ostium secundum defects
- Transesophageal echocardiography
- Defining size of defects
- Diagnosis of sinus venosus defects
- Assessment of joined congenital anomalies
- Assessment of joined abnormalities (mitral valve prolapse)

- Doppler flow echocardiography for measuring shunt volume, shunt ratios, and pulmonary artery pressures

An echocardiogram might reveal right ventricular hypertrophy. Prolongation of the PR interval is commonly seen in all types of atrial septal defects.

Management of Pediatric Atrial Septal Defects

An important question concerning ASD is whether the treatment itself is needed or the defect should be left to heal on its own.

Medical treatment of atrial septal defects is not helpful. All patients with atrial septal defects will need a surgical intervention to close the defect except for ostium secundum defects which are known to close spontaneously in a significant number of cases.

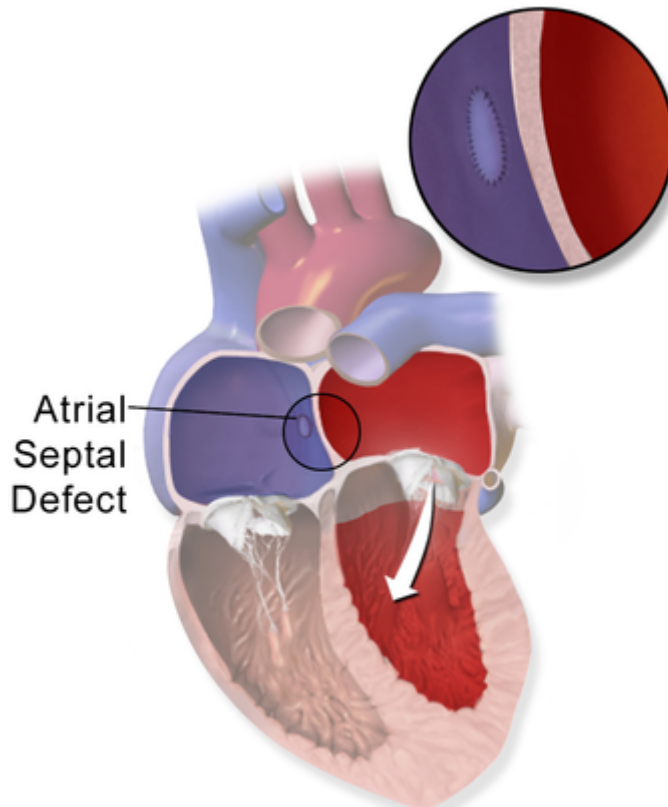
Patients with atrial septal defects who develop congestive heart failure should receive diuretics and possibly digoxin. Atrial fibrillation is rarely seen in children.

Prophylaxis against bacterial endocarditis is not needed in patients with atrial septal defects. Patients who undergo surgical or percutaneous repair of their atrial septal defect should receive antibiotic prophylaxis for a period of six months. Additionally, patients who are found to have residual persistent left to right shunting after surgical intervention should receive antibiotic prophylaxis against bacterial endocarditis indefinitely.

Surgical closure of atrial septal defects is usually reserved for patients with significant left-to-right shunting. This can be confirmed by a pulmonary-to-systemic flow ratio of more than 1.5:1 or by significant enlargement of the right atrium and ventricle on echocardiography. Another important indication for surgical closure of atrial septal defects is the development of congestive heart failure.

Percutaneous minimally invasive catheterization-based techniques have been developed for the closure of atrial septal defects. These procedures carry a very low mortality rate and are usually very successful. Because part of the pericardium is used to form the patch used to close the atrial septal defect, up to one-third of the patients might develop the post-pericardiotomy syndrome.

Patients with the post-pericardiotomy syndrome have fluid accumulation in the pericardial space after pericardiotomy. Catheter-based approaches for the closure of atrial septal defects are usually reserved for children aged between 4 and 6 years of age who have a significant hemodynamic instability because of atrial septal defect.



Patch Closure for Atrial Septal Defect

[Image:](#) "Atrial Septal Defect (Patch Closure)." . By Blausen.com staff (2014).
"Medical gallery of Blausen Medical 2014". Wikijournal of Medicine 1 (2).
DOI:10.15347/wjm/2014.010. ISSN 2002-4436.. License: [CC BY 3.0](#)

1. Indications for intervention are not uniform, but most of the recommendations include the following:

- ASD patients with echocardiographic evidence of right-sided cardiac volume loading
- Symptomatic ASD patients (in the first place exercise related)
- ASD patients with exercise-related cyanosis, although not accompanied by pulmonary hypertension
- ASD patients with an anamnesis of paradoxical thromboembolic episode
- May be used as a prophylaxis procedure in non-cardiac, high risk paradoxical embolization , intervention in ASD patients (joint replacement or similar)

2. An absolute contraindication is:

- Pulmonary hypertension, accompanied by R-to-L shunt at rest or pulmonary vascular resistance of > 14 Wood units

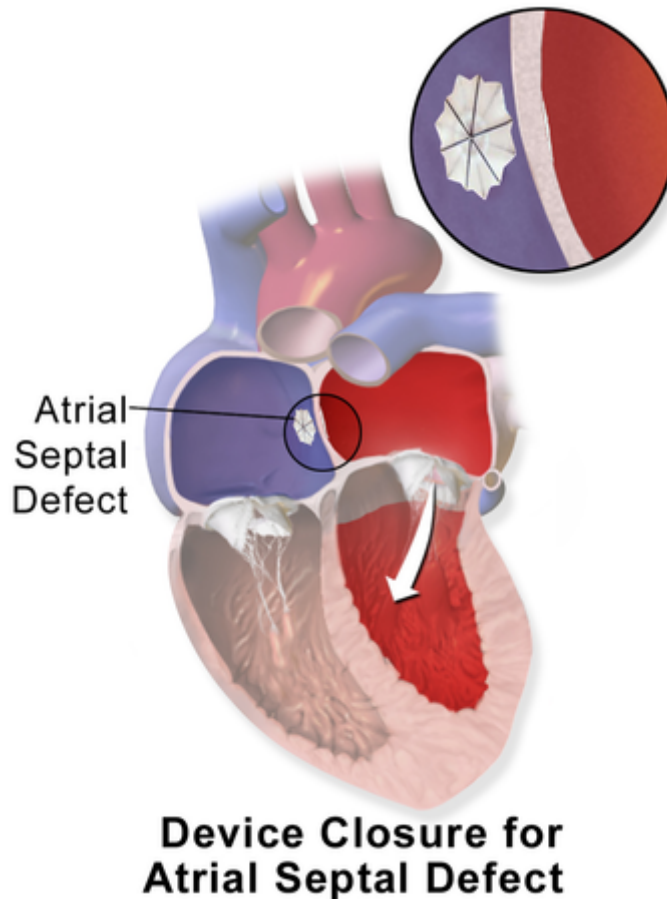
3. Relative contraindications present:

- Patients with intermediate degrees of pulmonary hypertension

- Patients with bidirectional shunt
- Patients with predominantly L-to-R shunts

4. Regarding the type of intervention:

- Open surgical approach — since 1954
- Transcatheter closure — since 1976 (the most commonly used device being Amplatz septal occlude)
- New techniques — like robotic assistance



[Image:](#) "Device Closure for Atrial Septal Defect." . By Blausen.com staff (2014).
"Medical gallery of Blausen Medical 2014". Wikijournal of Medicine 1 (2).
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Complications of Pediatric Atrial Septal Defects

Complications in symptomatic ASD patients usually do not develop until adulthood, approximately around age 30. Rarely complications occur in infants and children.

Potential ASD's complications include:

- Right heart failure
- [Arrhythmias](#)
- [Stroke](#)
- [Pulmonary hypertension](#)

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