

### What is an atrial septal defect (ASD)?

The heart has four chambers: two receiving chambers called right and left atria and two pumping chambers called right and left ventricles. The atrial septum is the wall that separates the left and right atria. If there is a hole in the atrial septum, it is called an atrial septal defect (ASD). Some of the blood that should flow into the left ventricle (or lower pumping chamber) from the left atrium now flows into the right atrium through the ASD. In turn, more blood goes to the right side of the heart and back to the lungs rather than out to the body. This is referred to as a “shunt.”

There are four types of ASDs. The most common is the secundum ASD (75%). Others are primum (20%), sinus venosus (4%) and coronary sinus (<1%) ASD. Each is unique not only in its location and size but in how it “acts” and how it is treated.

When the ASD or shunt is small, only a little blood flows from one atrium to the other. These often don’t require treatment if heart size or function is not affected. Many small ASDs may close on their own as the heart grows during childhood. On the other hand, medium and large sized ASDs are less likely to close on their own and can cause complications if not closed.

### How do ASDs develop?

The heart starts out as a single tube in the developing fetus. As this tube loops, the two top tubes lie side-by-side and a wall (septum) develops. The atrial septum forms between the fourth week and fifth week of gestation. If this wall does not grow completely, a hole remains.

### What causes an ASD to develop?

No one knows exactly why an ASD forms. Mostly, they occur sporadically. Sometimes they are familial. It is thought that genetics and environmental factors may play a combined role.

### How common are ASDs?

ASDs are one of the most common congenital heart defects (CHDs). According to the Centers for Disease Control and Prevention (CDC) about 2,000 babies in the United States are born with an ASD every year. This is between 5% and 10% of all CHDs. They are twice as common in women as men and are the most frequently diagnosed CHD in adults.

### What other heart problems can occur with an ASD?

An ASD can occur alone or in association with other heart defects, such as anomalous pulmonary veins, pulmonary stenosis, tetralogy of Fallot and ventricular septal defect. If you are born with a more complex defect in addition to an ASD, the more complex defect is your primary diagnosis.

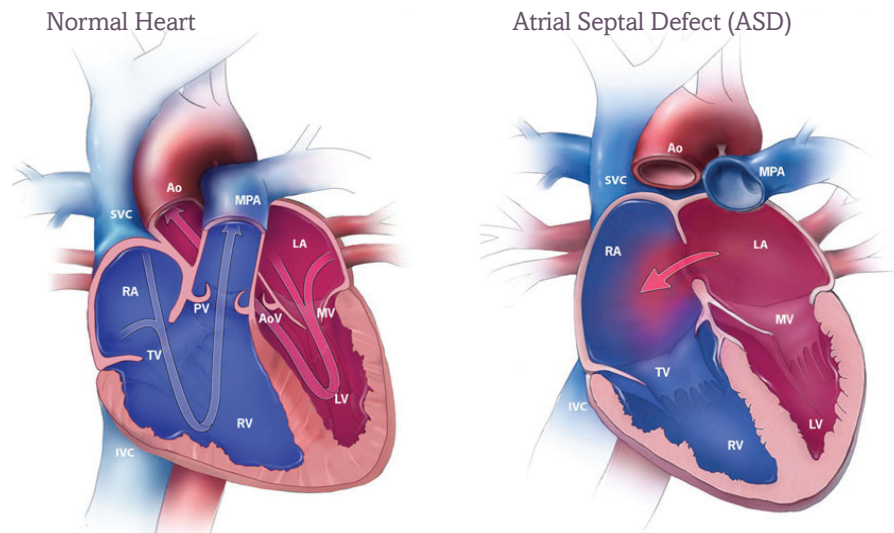
### How is an ASD diagnosed?

If the ASD is large, your doctor may hear an abnormal heart murmur when listening to your heart. In smaller ASDs, these may not be heard. Because many individuals with uncorrected secundum ASDs do not have significant symptoms, the ASD may not be found until later in childhood or adulthood.

The most common diagnostic test used to confirm an ASD is an echocardiogram (echo) or ultrasound of the heart.

RA – Right Atrium  
RV – Right Ventricle  
LA – Left Atrium  
LV – Left Ventricle  
SVC – Superior Vena Cava  
IVC – Inferior Vena Cava  
MPA – Main Pulmonary Artery  
Ao – Aorta  
AoV – Aortic Valve  
TV – Tricuspid Valve  
MV – Mitral Valve  
PV – Pulmonary Valve

Image courtesy of the Centers for Disease Control and Prevention, National Center on Birth Defects and Developmental Disabilities



Other tests your doctor may want you to have might include:

- Chest X-ray
- Electrocardiogram (EKG)
- Cardiac Magnetic Resonance Imaging (MRI)
- Transesophageal Echo (TEE)
- Heart Catheterization

### **What types of symptoms should I look for?**

In adulthood, symptoms are often subtle. You may have decreased exercise tolerance, shortness of breath, increased fatigue, irregular heartbeats, palpitations and/or fainting. If left untreated, you might be at increased risk for stroke, heart failure and/or increased pressure in the lungs, known as pulmonary hypertension (PH).

### **If I have an ASD, can I get pregnant?**

All women who have an ASD, repaired or not, small or large, should see an adult congenital heart disease (ACHD) doctor before getting pregnant to assess heart function and risk. While most women with a small or repaired ASD will do well during pregnancy, there is some risk. Those with a larger unrepaired ASD, and/or additional heart or lung problems, have an increased risk for complications during pregnancy and after delivery. Remaining in the care of experts in heart defects and a high-risk pregnancy obstetrician both prior to and during the entire pregnancy is important.

### **If I have an ASD, do my children have a higher risk of having a heart defect?**

Adults with CHDs are more likely than the general population to have a child with a heart defect. The risk is higher if the mother is the parent with a CHD and if there is a sibling with CHD. The defects may be the same or different than the parent's defect.

### **When do you decide to repair?**

Up to 40% of secundum ASDs close on their own by adulthood. If the ASD remains but is small, closure is usually not recommended. However, if you develop symptoms such as fatigue, difficulty breathing, stroke, atrial fibrillation, or PH, or if your right ventricle becomes enlarged, repair may be considered. If you have severe pulmonary hypertension due to Eisenmenger syndrome, closure may not be recommended.

### **How are ASDs closed?**

In 1953, Dr. John H. Gibbon successfully closed an ASD using open heart surgery and a heart-lung machine. Since then, ASDs have been closed using different materials and approaches. Until the early 1990s, all ASDs were closed with open heart surgery. Now, device closure through a cardiac catheterization has become the first choice to repair secundum ASDs if certain criteria are met. If the secundum ASD is too large or if the patient has any of the other types of ASD, surgical closure is recommended. Surgical closure

involves opening the atrium and closing the defect with a suture, patch or plug. Your surgeon will work with you to decide the best method to close your ASD.

### **Can I exercise?**

It is important to ask your ACHD doctor before starting any new physical activity routine. People with a small or repaired ASD, however, usually have no exercise limitations. If you have other problems, such as an irregular heartbeat, PH, heart failure, or Eisenmenger syndrome, ask your ACHD doctor what type of exercise is safe for you.

### **What are the long-term outcomes for adults with unrepaired ASD?**

Adults with small ASDs that do not affect heart function usually don't require treatment. It is still important to see an ACHD doctor at least once to make sure nothing has changed. On the other hand, if the hole between the two sides is too big, blood then flows from the left side of the heart into the right side. As a result, extra blood is pumped to the lungs, making the heart and lungs work harder and less efficiently. When this happens, there is a greater risk of developing other heart, lung, and blood problems, including:

- An enlarged right heart, which can cause right heart failure
- Irregular and rapid heartbeats, such as atrial fibrillation, particularly in the atria
- Stroke
- Damage to the lung arteries, causing PH
- Eisenmenger syndrome
- Leaking tricuspid and mitral valves

### **What are the long-term outcomes for adults with repaired ASD?**

With early diagnosis and repair of an ASD, the outcome is generally excellent. Those whose ASD is diagnosed and closed in adulthood and who have normal pulmonary artery pressures are also expected to have a good long-term outcome. They do, however, have an associated risk of developing atrial arrhythmias because of scars at the area of closure. In a small number, re-operation may be needed if a leak occurs at the closure area. Other risks that can occur at a later time include the chance of developing heart failure or PH.

### **What kind of heart care is recommended for adults with repaired and unrepaired ASD?**

Experts recommend that all adults who have an unrepaired ASD be followed regularly throughout their life by an ACHD specialist. Those whose ASD was closed, whether as a child or adult, need periodic heart checks; as adults, they should be seen at least once at an ACHD center. This is to make sure that the diagnosis is correct and the repair is still working. Further need for follow-up will be determined by the ACHD cardiologist.

For their work on this article, ACHA thanks **Naser M. Ammash, MD**, Mayo Clinic, Rochester, MN and **Ali Zaidi, MD**, Montefiore Adult Congenital Heart Disease Program (MAAtCH), Bronx, NY.