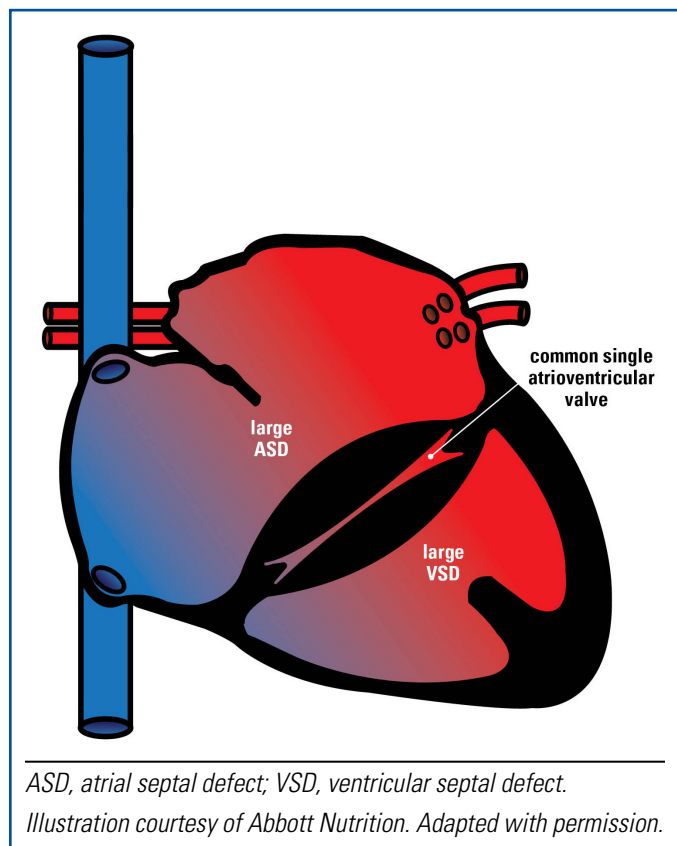




Cardiac Defects: Atrioventricular Canal Defects



An *atrioventricular canal defect* is a problem in the part of the heart that connects the upper chambers (atria) to the lower chambers (ventricles). There are two types of atrioventricular canal defects: complete and partial.

Complete Atrioventricular Canal (CAVC)

Complete atrioventricular canal (CAVC) defect is a severe defect in which there is a large hole in the tissue (the septum) that separates the left and right sides of the heart. The hole is in the center of the heart, where the upper chambers (the atria) and the lower chambers (the ventricles) meet.

As the heart formed abnormally, with this large hole, the valves that separate the upper and lower chambers also developed abnormally. In a normal heart, two valves separate the upper and lower chambers of the heart: the tricuspid valve separates the right chambers and the mitral

valve the left. In a child with CAVC defect, there is one large valve that may not close correctly.

As a result of the abnormal passageway between the two sides of the heart, blood from both sides mix and too much blood circulates back to the lungs before it travels through the body. This means the heart works harder than it should have to, and it will become enlarged and damaged if the problems aren't repaired.

Partial Atrioventricular Canal Defects

A *partial atrioventricular canal defect* is the less severe form of this heart defect. The hole does not extend between the lower chambers of the heart and the valves are better formed. Usually it is necessary only to close the hole between the upper chambers (this hole is called an *atrial septal defect*, or ASD) and to do a minor repair of the mitral valve. Partial atrioventricular canal is also called *atrioventricular septal defect*, or AVSD.

What are the symptoms of atrioventricular canal defects?

In CAVC defect, the following symptoms may be present within several days or weeks of birth:

- blue or purple tint to lips, skin, and nails (cyanosis)
- difficulty breathing
- poor weight gain and growth
- heart murmur—the heart sounds abnormal when a doctor listens with a stethoscope.

Partial atrioventricular canal defects cause fewer symptoms and sometimes aren't diagnosed until the child reaches his or her 20s or 30s and begins to experience irregular heartbeat (arrhythmia), leaky valves, or other effects.

How are atrioventricular canal defects diagnosed?

The healthcare provider who evaluates the newborn in the hospital might make the diagnosis, or a primary care provider might notice a murmur and other symptoms and refer the baby to a cardiologist.



Diagnosis of atrioventricular canal defects may require some or all of these tests:

- echocardiogram—sound waves create an image of the heart
- electrocardiogram (ECG)—a record of the electrical activity of the heart
- chest X ray
- pulse oximetry—a noninvasive way to monitor the oxygen content of the blood
- cardiac catheterization—a thin tube is inserted into the heart through a vein and/or artery in either the leg or through the umbilicus (“belly button”)
- cardiac MRI—a three-dimensional image shows the heart’s abnormalities.

Sometimes a complete atrioventricular canal defect is diagnosed on a fetal ultrasound or echocardiogram. Your baby’s providers can prepare a plan for delivery and care immediately after birth.

Complete atrioventricular canal defects often occur in children with Down syndrome.

What are the treatment options for atrioventricular canal defects?

Complete atrioventricular canal defects require surgery, usually within the first 2 or 3 months of life. The surgeon will close the large hole with one or two patches. The patches are stitched into the heart muscle. As the child grows, the tissue grows over the patches.

The surgeon will also separate the single large valve into two valves and will reconstruct the valves so they are as close to normal as possible, depending on the child’s heart anatomy.

Partial atrioventricular canal defects also require surgery, whether it is diagnosed in childhood or adulthood. The

surgeon will patch or stitch the atrial septal defect closed and will repair the mitral valve or replace it with either an artificial valve or a valve from a donated organ.

After surgery, patients recover in an intensive care unit as they improve.

What kind of follow-up care is required for atrioventricular canal defects?

Through Age 18

A child who has had surgical repair of an atrioventricular canal defect will require lifelong care by a cardiologist. Most children recover completely and won’t need additional surgery or catheterization procedures.

Pediatric cardiologists follow patients until they are young adults, coordinating care with the primary care provider. Patients will need to carefully follow providers’ advice, including staying on any medications prescribed and, in some cases, limiting exercise.

Sometimes children with an atrioventricular canal defect experience heart problems later in life, including irregular heartbeat (arrhythmia) and leaky or narrowing valves. Medicine, additional surgery, or cardiac catheterization may be required.

Into Adulthood

Pediatric cardiologists will help patients transition care to an adult congenital heart disease specialist. Because of enormous strides in medicine and technology, today most children born with atrioventricular canal defects go on to lead productive lives as adults.

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