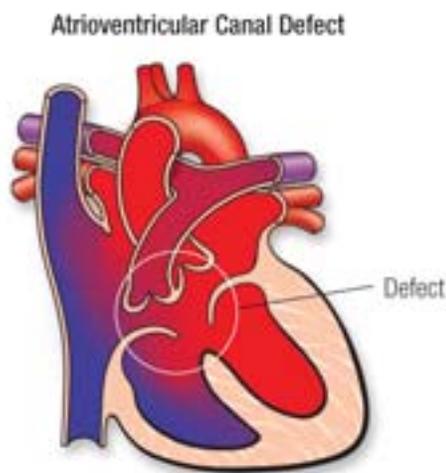


## Complete Atrioventricular Canal (CAVC)

### What is it?

Atrioventricular (AV) canal defect is a large hole in the center of the heart. It's located where the wall (septum) between the upper chambers (atria) joins the wall between the lower chambers (ventricles). Many terms are used to describe this heart defect, including atrioventricular (AV) canal, complete AV canal, complete common AV canal, and endocardial cushion defect. Essentially, they all describe a similar heart problem. It's a combination of:

1. a hole in the wall dividing the heart's upper chambers (atrial septal defect),
2. a hole in the wall separating the heart's lower chambers (ventricular septal defect) and
3. abnormalities of the tricuspid and mitral valves.



Some patients with AV canal defects have a small or no opening between the bottom chambers of the heart. They can present in adulthood with findings similar to patients with atrial septal defects; this can be referred to as an “ostium primum” atrial septal defect. Even less commonly, only the hole between the lower chambers is present.

### What causes it?

In most cases, the cause isn't known. It's a very common type of heart defect in children with a chromosome problem, Trisomy 21 (Down syndrome). Some patients can have other heart defects along with AV canal.

### How does it affect the heart?

Normally, the left side of the heart only pumps blood to the body, and the heart's right side only pumps blood to the lungs. In a child with AV canal defect, blood can travel across the holes from the left heart chambers to the right heart chambers and out into the lung arteries. The extra blood being pumped into the lung arteries makes the heart and lungs work harder, and the lungs can become congested. Sometimes there's leakiness (regurgitation) of the abnormal single valve. This may add to the heart failure symptoms.

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### How does the AV canal defect affect patients?

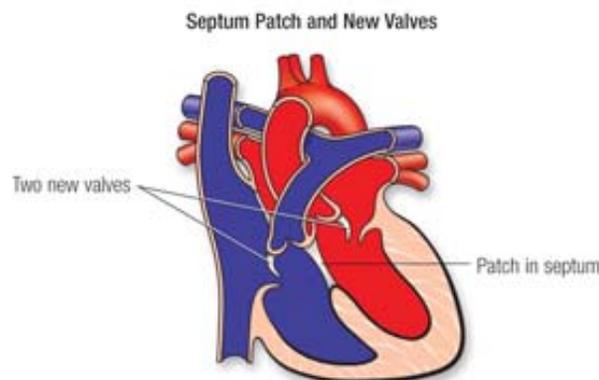
High pressure may occur in the blood vessels in the lungs that can lead to permanent damage with pulmonary hypertension that persists into adulthood.

Many adults who have not had previous repair have pulmonary hypertension (see Eisenmenger's syndrome). This complication is more common than in patients with an ASD or a VSD. Even in adults without Eisenmenger's syndrome, symptoms including shortness of breath, intolerance to exercise and palpitations are common. On physical examinations, murmurs due to the blood flow across the defects and due to the valve leak are common.

### If my AV canal defect was closed in childhood, what was done and what can I expect?

The surgery to fix these defects involves patching the ASD and the VSD and repairing the heart valve. To repair the valve, the surgeon divides the single valve between the heart's upper and lower chambers and makes two separate valves. These are made as close to normal valves as possible. It's possible that a temporary operation to relieve symptoms and high pressure in the lungs may have been performed before the definitive operation. This procedure (pulmonary artery banding) narrows the pulmonary artery to reduce the blood flow to the lungs. When the child was older, an operation was done to remove the band and fix the AV canal defect with open-heart surgery. Unlike some other types of septal defects, the AV canal defect never closes on its own.

Surgical repair of an AV canal usually restores blood circulation to normal. For many patients, the long-term outlook is good, and no medicines or additional surgery are needed. Because this is a more complicated congenital heart defect, late problems in adults are more common than after an ASD or VSD is closed. As the child grows, the repair may partially break down leading to patch leaks, valve leakage and narrowing of the blood flow channel to the body. These problems may increase the workload of the heart and cause symptoms.



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### What if the AV canal defect is still present? Should it be repaired in adulthood?

The decision to repair an AV canal defect in adulthood is complicated. It depends on the pressures in the lung and the heart's pumping function. However, when the pressures aren't too high and the pump function is good, these defects can be repaired and adult patients are likely to improve. A heart catheterization is almost always required to know whether the defect should be closed. These defects can't be closed or repaired in the catheterization laboratory, however, because of their location and the need to fix the heart valves.

### Problems You May Have

Problems in patients with repaired AV canals depend on whether there are patch leaks and whether there's a lot of valve regurgitation. Shortness of breath, inability to exercise and swelling in the legs are all signs of heart failure. Abnormal heart rhythms may cause palpitations (skipped or rapid heartbeats) and, rarely, fainting. Some patients may need a pacemaker after the repair if the electrical system has been damaged.

Patients with unrepaired AV canal are often blue. Because of valve leaks on the heart's left side, they're more likely to have heart failure than other patients with Eisenmenger's syndrome, due to ASDs and VSDs.

### Ongoing Care:

#### What will I need in the future?

An adult with a repaired or unrepaired AV canal defect must be examined regularly by a cardiologist with experience in adult congenital heart disease. The frequency of the visits depends on the extent of problems with the repair, the presence of abnormal heart rhythms and pulmonary hypertension. In general, you should visit the cardiologist at least once a year. You should also consult a cardiologist with expertise in care of adult congenital heart disease if you're undergoing any type of non-heart surgery or invasive procedure or thinking about heart surgery.

### Medical

Heart failure medications may be needed, especially in patients with valve regurgitation, to help their heart pump better and/or lower blood pressure. Patients with pulmonary hypertension may also require medications. Your cardiologist can monitor you with noninvasive tests if needed. These include electrocardiograms, Holter monitors, exercise stress tests and echocardiograms. When further surgery is contemplated, a heart catheterization is almost always needed.

#### What activities can I do?

If the AV canal defect has been closed with surgery, you may not need any special precautions and may be able to participate in normal activities without increased risk. However, patients with heart failure and pulmonary hypertension may need some restrictions.

## **Complete Atrioventricular Canal (CAVC)**

### **Endocarditis Prevention**

Patients with AV canal defects may risk an infection of the heart's inside lining or valves (endocarditis) before and after surgery. Lifelong endocarditis prophylaxis is recommended.

### **Pregnancy**

If after surgery you have no significant residual (leftover) problems, the risk from pregnancy is low. If there are problems like a leaking valve or irregular heart rhythms, you may be at increased risk for complications of pregnancy. If you have heart failure or pulmonary hypertension, pregnancy isn't recommended. Women with unrepaired AV canal defects or who have leftover problems should talk to their cardiologist before deciding to get pregnant. Pregnant women with repaired AV canal defects who are free of significant problems may not require high risk obstetrical care. In contrast those unrepaired AV canal defects, significant valve leaks or pulmonary hypertension.

### **Will you need more surgery?**

The function of the repaired valves is a long-term concern. Some patients will need their valve replaced with a mechanical one when they get older. It's rare that the valve can be further repaired. Other patients may need more surgery to close patch leaks.