What Is It?

This is a combination of two defects:

1) An Atrial Septal Defect (ASD), or hole in the septum (muscle wall) that separates the right and left upper chambers of the heart (atria). In PAVC, the atrial septal defect forms in the lower part of the atrial septum, which is known as an ostium primum type defect.

2) A malformation of the Mitral Valve (MV), the one-way valve between the left atrium and the left ventricle, or main pumping chamber of the heart. This consists of a cleft or notch in the "flaps" or leaflets of the valve, preventing full closure of the valve.
What Are Its Effects?

Children with this defect usually have a heart murmur and have difficulty feeding and breathing and grow poorly.

The atrial septal defect allows the mixing of oxygen-rich blood from the left atrium with oxygen-poor blood in the right atrium. This is known as a left to right shunt. The mixed blood in the right atrium is pumped to the lungs via the right ventricle, reducing the efficiency of the circulatory system. This may lead to heart failure with congestion of the lungs.

Eventually, the atrial septal defect will cause the enlargement (dilatation) of the right atrium and right ventricle, which may lead to irregular atrial pumping (arrhythmia) and/or right ventricular dysfunction.

The malformed mitral valve allows the leakage of blood between the left atrium and the left ventricle. The amount of leakage depends on the degree of deformation of the mitral valve. With time, a cleft mitral valve can result in the enlargement (dilatation) of the left atrium.

How Is It Treated?

In the treatment of Atrioventricular Canal Defect - Partial, the Mitral Valve is repaired by suturing the cleft or slit so that the valve no longer leaks (see illustrations).
The atrial septal defect (ASD) is closed by suturing a patch (pink oval in the illustrations below) made of pericardium (the membrane that covers the heart) or a synthetic material such as Dacron or Teflon over the hole.

Postoperative recovery is usually rapid, requiring a hospital stay off from four days to one week.