

Pulmonary Atresia (PA)

With Ventricular Septal Defect and MAPCAs Single Stage Repair

When PA occurs the pulmonary valve does not develop and there is a narrowing beneath it. The main pulmonary artery and the pulmonary branch arteries are very small. There is a hole between the bottom two chambers of the heart (ventricular septal defect or VSD). And, there may or may not be a connecting blood vessel from the aorta to the pulmonary artery (patent ductus arteriosus or PDA).

When the main pulmonary artery and/or its branches are small, it is hard for blood to get to the lungs to pick up oxygen. The pulmonary artery blood flow comes from major aortopulmonary collateral arteries (MAPCAs). These MAPCAs arise from the descending aorta (the part of the aorta that carries blood and oxygen down to the body, legs and feet).

The corrective surgery is aimed at directing the blood flow from the bottom right chamber of the heart to the branch arteries going to the lungs. The single stage operation includes these steps:

1. The MAPCAs are disconnected from the aorta. The main pulmonary artery is removed. The branch pulmonary arteries are disconnected from the main artery when it is removed. The MAPCAs are then opened and sewn into the pulmonary artery branches to form a larger vessel. This procedure is called unifocalization of the MAPCAs. (Unifocalization means bringing two vessels together to make them one larger vessel.)
2. The VSD is closed using a patch of material called Dacron®.
3. A conduit is placed in the right ventricle and sewn into the branch pulmonary arteries and MAPCAs. Blood can now flow from the right ventricle through the conduit into both the pulmonary branch arteries and MAPCAs and into the lungs.

This surgery is done through a median sternotomy (chest) incision.

