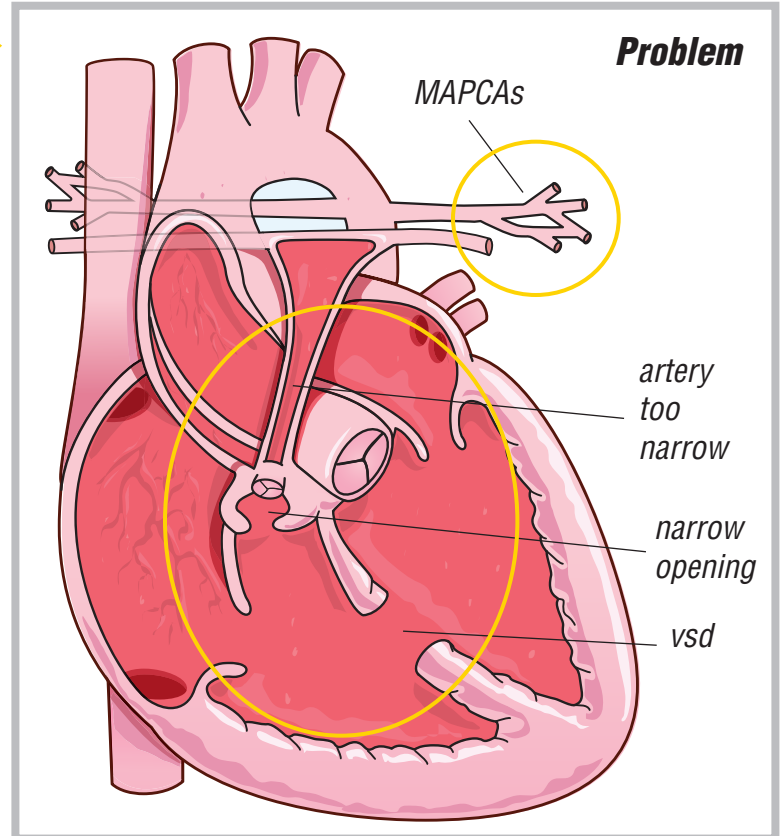


Pulmonary Atresia (PA)

With Ventricular Septal Defect and MAPCAs

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).



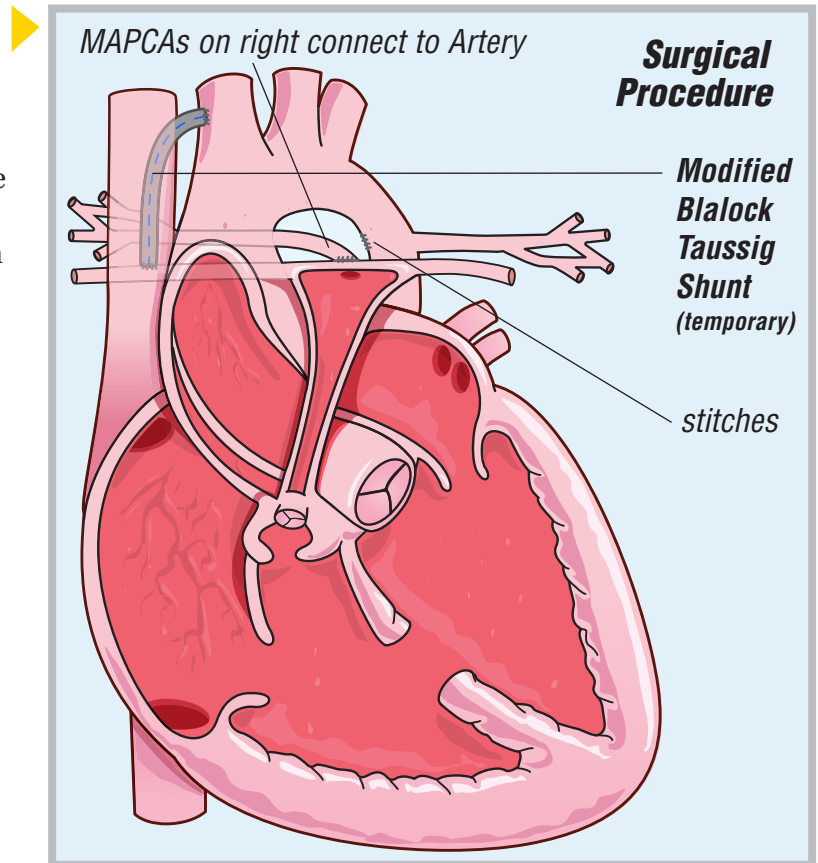
The corrective surgery is aimed at directing the blood flow from the bottom right chamber of the heart to the branch arteries going to the lungs. The repair includes three (or two) staged operations:

Pulmonary Atresia (PA)

With Ventricular Septal Defect and MAPCAs Stage 1

Stage 1 -

The MAPCAs on the right side are disconnected from the right side of the aorta. They are then opened and sewn into the right pulmonary artery branch 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.) A small tube (made either from a blood vessel of the infant or artificial material) is placed from the right subclavian artery into the right branch pulmonary artery (modified Blalock Taussig Shunt). The tube (shunt) allows blood to enter the lungs at all times. The shunt is placed through a thoracotomy (side) incision.



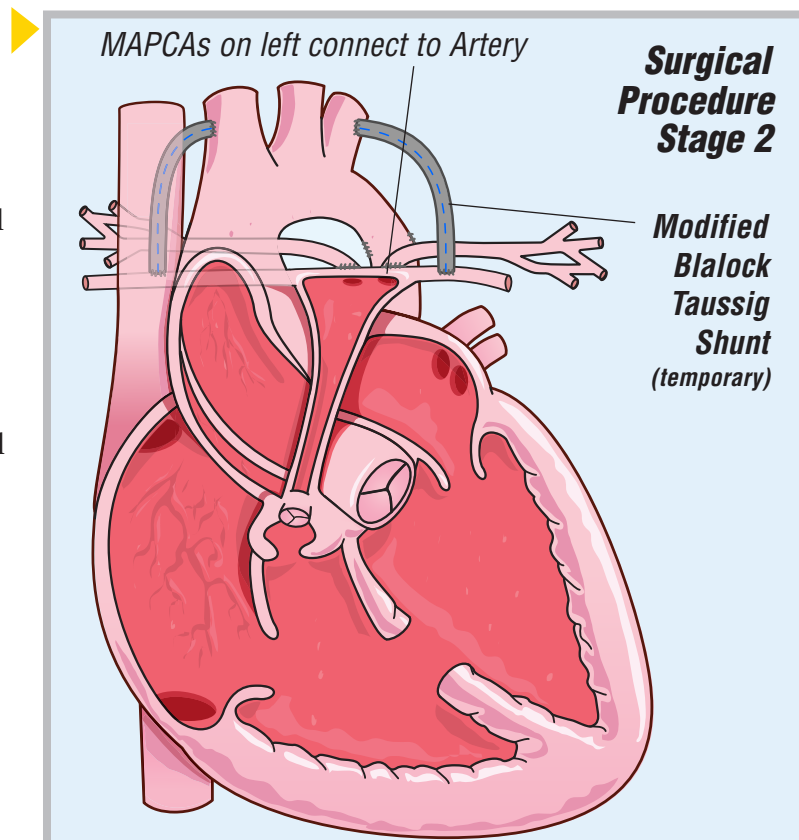
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Pulmonary Atresia (PA)

With Ventricular Septal Defect and MAPCAs Stage 2 and 3

Stage 2

The MAPCAs on the left side are disconnected from the left side of the aorta. They are then opened and sewn into the left pulmonary artery branch as in stage 1. A small tube (made either from a blood vessel of the infant or artificial material) is placed from the left subclavian artery into the left branch pulmonary artery (modified Blalock Taussig Shunt). The tube (shunt) allows more blood to enter the lungs at all times. The shunt is placed through a thoracotomy (side) incision. (This operation is sometimes done at the same time as Stage 1 and at other times is not needed.)



Stage 3

The main pulmonary artery is removed. The branch pulmonary arteries are disconnected from the main artery when it is removed. A conduit is made to attach the right ventricle to the branch pulmonary arteries and MAPCAs. Blood can now flow from the right ventricle through the conduit into both branch pulmonary arteries and MAPCAs and into the lungs. The VSD is closed using a patch of material called Dacron®. The shunts from Stage 1 and Stage 2 are removed. This surgery is done through a median sternotomy (chest) incision.

