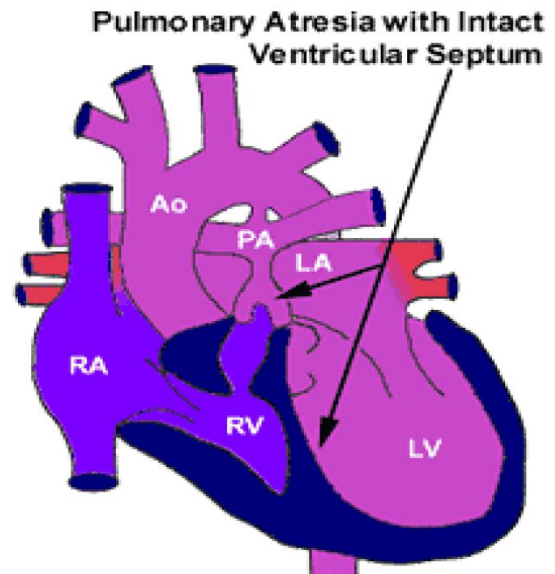
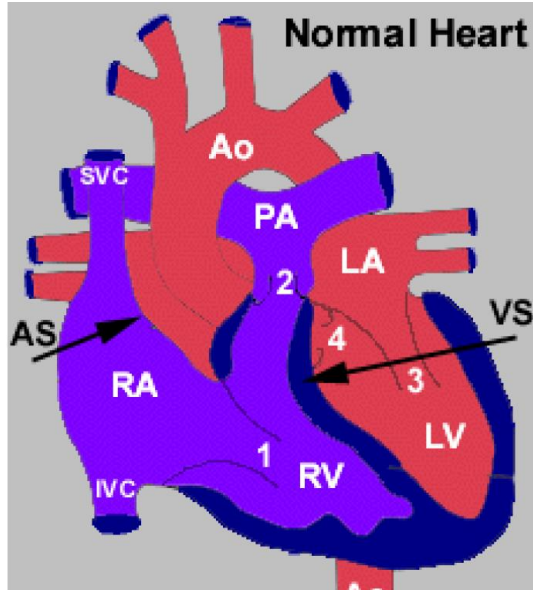


## Pulmonary Atresia With Intact Ventricular Septum



### Anatomy:

The term “atresia” indicates failure of the pulmonary valve or pulmonary trunk vessel to develop. Therefore, there is no connection between the RV and pulmonary artery. The pulmonary valve annulus may be very small, and the main pulmonary artery may be absent or rudimentary. The right and left pulmonary arteries may be of normal size, or they may be extremely small. When there is pulmonary atresia with intact ventricular septum, the RV is usually extremely small, thick-walled and the tricuspid valve is often stenotic, small or deformed.

### Physiology:

If no VSD is present, systemic venous blood that enters the right heart quickly fills the RV but has no outflow path.

The increase in the RA pressure opens the foramen ovale, so that systemic venous blood flows from the right to the left atrium and mixes with the pulmonary venous blood. The mixed blood enters the LV and is ejected into the aorta. The entire pulmonary blood flow is supplied by the PDA. There must be a PDA to maintain mixing. Upon diagnosis, PGE is started to maintain ductal patency. If flow is not adequate, a Rashkind balloon septostomy may be performed during cardiac cath to enable better flow from the right to left atrium.

### Surgery: *Palliative*

BT Shunt, followed by Glenn. The Glenn (or hemiFontan) generally connects the SVC to the right pulmonary artery. The Glenn is followed by the Fontan (more details in the tricuspid atresia section). In the Fontan, the IVC is connected to the pulmonary artery. There are many versions of the Fontan procedure that all generally serve to provide continuity between the systemic venous return and the pulmonary artery.