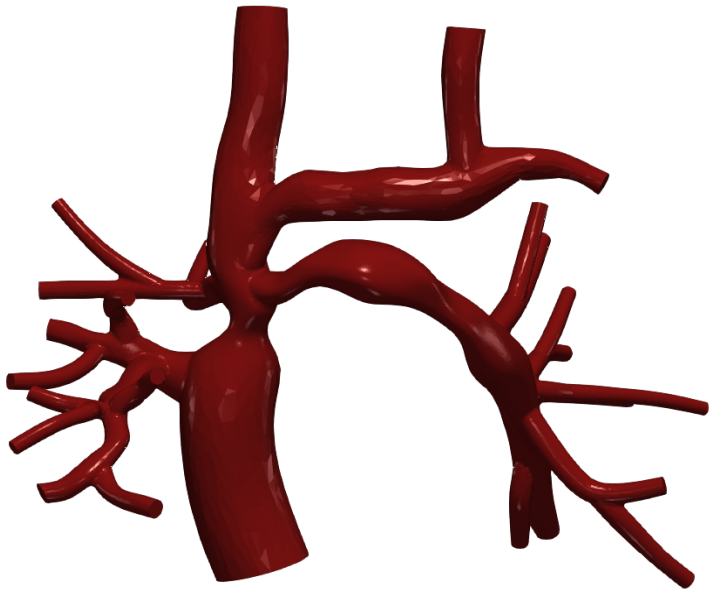
Vascular Model Repository Specifications Document



0056_H_PULMFON_TAT

Legacy Name: 0065_0001

Model added: 27 Dec 2021

Species	Human
Anatomy	Pulmonary Fontan
Disease	Tricuspid Atresia
Procedure	Fontan

Clinical Significance and Background

Pulmonary Fontan

The pulmonary arteries are the blood vessels responsible for transporting deoxygenated blood to the lungs to perform respiration. A normal pulmonary anatomy involves the main pulmonary artery (MPA) trunk leaving the right ventricle of the heart which then branches off into the left and right pulmonary arteries which then continue to fractally branch towards the lungs. However, after the Fontan procedure (an open-heart surgery done on babies with congenital heart defects) the anatomy of the pulmonary system is slightly modified. The main pulmonary artery trunk is not present. Instead, the superior vena cava (SVC) and inferior vena cava (IVC) are directly connected to the left and right pulmonary arteries in a 4-way intersection configuration. This is done so that deoxygenated blood coming from the upper body (through the SVC) and deoxygenated blood coming from the lower body (through the IVC) flow directly to the lungs without passing through the heart to allow for the heart to focus solely on pumping oxygenated blood from the lungs to the rest of the body.

Tricuspid Atresia

Tricuspid atresia is a birth defect of the tricuspid valve, which is the valve that controls blood flow from the right atrium (upper right chamber of the heart) to the right ventricle (lower right chamber of the heart). Tricuspid atresia occurs when this valve does not form at all, and no blood can go from the right atrium through the right ventricle to the lungs for oxygen.

In tricuspid atresia, since blood cannot directly flow from the right atrium to the right ventricle, blood must use other routes to bypass the unformed tricuspid valve. Babies born with tricuspid atresia often also have an atrial septal defect, which is a hole between the right and left atria, or a ventricular septal defect, which is a hole between the right and left ventricles. These defects allow oxygen-rich blood to mix with oxygen-poor blood so that oxygen-rich blood has a way to get pumped to the rest of the body.

Fontan

The Fontan procedure is a type of open-heart surgery. Children who need this surgery usually have it when they are 18 - 36 months old. The Fontan procedure is done for children who are born with heart problems like hypoplastic left heart syndrome (HLHS),

tricuspid atresia, and double outlet right ventricle, and depending on the heart problem, children may need the Norwood procedure and Glenn procedure before the Fontan surgery.

After the Fontan procedure, the blood from the lower body goes directly to the lungs. The blood with high oxygen goes into the heart. This way the single ventricle only pumps blood to the body and only pumps blood with high oxygen to the body. There is no more mixing of oxygen-rich blood and oxygen-poor blood.

During the Fontan procedure, the surgeon first disconnects the inferior vena cava (IVC) from the heart and connects it to the pulmonary artery using a conduit (tube). Then, the surgeon makes a small hole between the conduit and the right atrium. This hole (or fenestration) lets some blood still flow back to the heart. It prevents too much blood from flowing to the lungs right away, so they have time to adjust. Doctors can close the fenestration later by doing a cardiac catheterization procedure.

Clinical Data

General Patient Data

Age (yrs)	5
Sex	Female

Specific Patient Data

BSA (m^2)	0.68
CI (L/min/m^2)	2.8
P IVC MP cath	11
P SVC MP cath	11
P LPA MP cath	7
P RPA MP cath	9

Notes

Paper patient "F". See paper for more details. See below for information on the image data.

Image Modality: CT

Image Type:	VTI
Image Source:	TLAB
Image Manufacturer:	SIEMENS

Publications

See the following publications which include the featured model for more details:

Marsden, A. L., Reddy, V. M., Shadden, S. C., Chan, F. P., Taylor, C. A., & Feinstein, J. A. (2010). A new multiparameter approach to computational simulation for Fontan assessment and redesign. Congenital Heart Disease, 5(2), 104-117. http://www.doi.org/10.1111/j.1747-0803.2010.00383.x

License