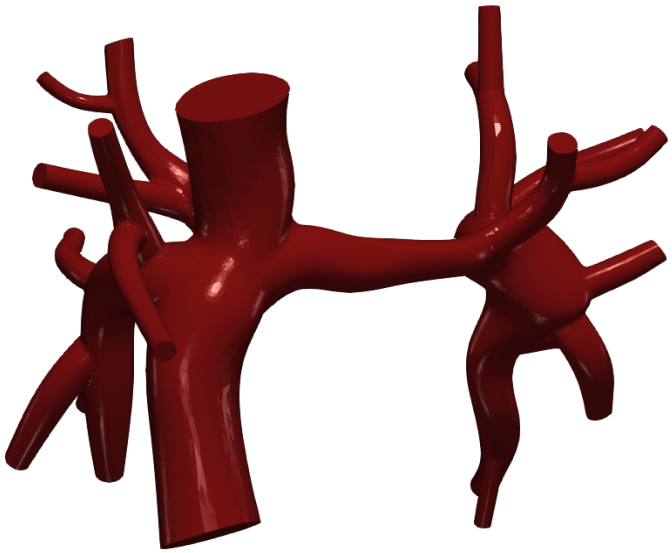
Vascular Model Repository Specifications Document



0064_0001

Species	Human
Anatomy	Pulmonary
Disease	Congenital Heart Disease Single Ventricle Defect Hypoplastic Left Heart Syndrome
Procedure	Fontan

Clinical Significance and Background

Pulmonary

The pulmonary circulation involves blood flowing from the right ventricle of the heart into the pulmonary arteries. From the pulmonary arteries, the blood then reaches the lungs, performs a gas exchange, and then continues to the pulmonary veins which then lead to the left atrium of the heart.

By definition, an artery is a blood vessel that carries blood away from the heart. This usually means arteries carry oxygenated blood to the rest of the body, but since the pulmonary arteries are transporting blood from the right side of the heart to the lungs to perform respiration, that makes the pulmonary arteries the only arteries in the body that actually carry deoxygenated blood. Similarly, the pulmonary veins, which carry blood that has been freshly oxygenated from the lungs back to the heart, are the only veins that actually carry oxygenated blood.

Congenital Heart Disease

Congenital heart disease is one or more problems with the heart's structure that exist since birth. Congenital means that you're born with the condition. Congenital heart disease in adults and children can change the way blood flows through the heart. Some types of congenital heart disease may be mild. But complex defects may cause life-threatening complications. However, advances in diagnosis and treatment continue to improve survival for those with congenital heart disease. People with congenital heart disease need lifelong medical care. Treatment may include regular checkups (watchful waiting), medications or surgery.

Single Ventricle Defect

A single ventricle defect (SVD) is a type of heart defect that a child is born with. It occurs when one of the two pumping chambers in the heart, called ventricles, isn't large enough or strong enough to work correctly. In some cases, the chamber might be missing a valve. Single ventricle defects are rare, affecting only about five out of 100,000 newborns. They are also one of the most complex heart problems, usually requiring at least one surgery. There are several types of single ventricle defects which include but are not limited to: tricuspid atresia, hypoplastic left heart syndrome (HLHS), mitral valve atresia (usually associated with HLHS), single left ventricle, double inlet left ventricle (DILV), double outlet right ventricle (DORV), pulmonary atresia with intact

ventricular septum (PA/IVS), Ebstein's anomaly, and atrioventricular canal defect (AV Canal).

Hypoplastic Left Heart Syndrome

In hypoplastic left heart syndrome, the left side of the heart can't properly supply blood to the body because the lower left chamber (left ventricle) is too small or in some cases doesn't exist. In addition, the valves on the left side of the heart (aortic valve and mitral valve) don't work properly, and the main artery leaving the heart (aorta) is smaller than normal.

For the first days of life, the right side of the heart can pump blood both to the lungs and to the rest of the body through a blood vessel that connects the pulmonary artery directly to the aorta (ductus arteriosus). The oxygen-rich blood returns to the right side of the heart through a natural opening (foramen ovale) between the right chambers of the heart (atria). When the foramen ovale and the ductus arteriosus are open, they are referred to as being "patent."

If the ductus arteriosus and the foramen ovale close - which they normally do after the first day or two of life - the right side of the heart has no way to pump blood out to the body. In babies with hypoplastic left heart syndrome, medication is necessary to keep these connections open and keep blood flowing to the body until heart surgery is performed.

Fontan

The Fontan procedure is a type of open-heart surgery. Children who need this surgery usually have it when they're 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)	6
Sex	Female

Specific Patient Data

BSA (m^2)	0.71
CI (L/min/m^2)	2.7
P IVC MP cath	9
P SVC MP cath	6
P LPA MP cath	6
P aorta SP cath	95
P aorta DP cath	63
P aorta MP cath	78

Notes

Paper patient "E". See DOI for more details. See below for information on the image data and boundary conditions associated with the model.

Image Modality: MR

Image Type: DICOM

Image Source: TLAB

Image Manufacturer: GE MEDICAL SYSTEMS

Boundary Conditions: Refer to boundary conditions in the SimVascular file.

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

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