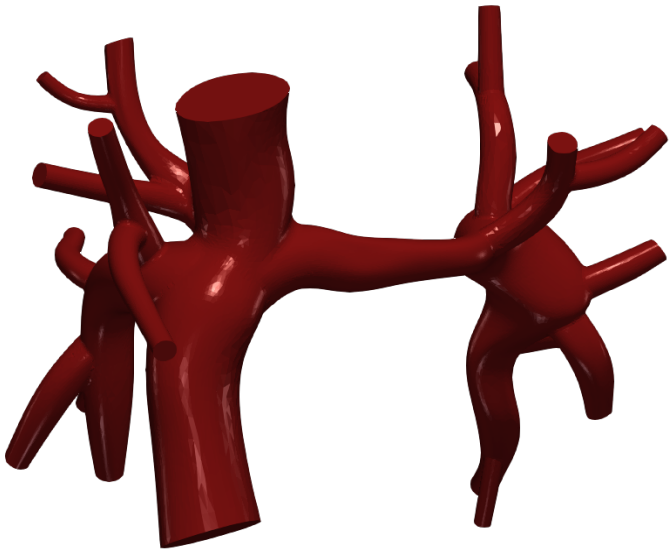
# Vascular Model Repository Specifications Document



# 0055_H_PULMFON_HLHS

Legacy Name: 0064_0001

Model added: 27 Dec 2021

| | |
|---|---|
| **Species** | Human |
| **Anatomy** | Pulmonary Fontan |
| **Disease** | Hypoplastic Left Heart Syndrome |
| **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.

## Hypoplastic Left Heart Syndrome

In hypoplastic left heart syndrome, the left side of the heart cannot properly supply blood to the body because the lower left chamber (left ventricle) is too small or in some cases does not exist. In addition, the valves on the left side of the heart (aortic valve and mitral valve) do not 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 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) | 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 paper for more details. See below for information on the image data.

| | |
|---|---|
| **Image Modality:** | MR |
| **Image Type:** | VTI |
| **Image Source:** | TLAB |
| **Image Manufacturer:** | GE MEDICAL SYSTEMS |

# 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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