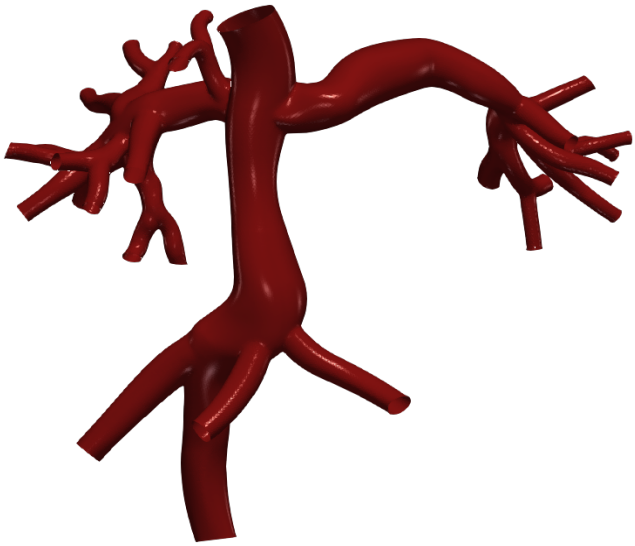
Vascular Model Repository Specifications Document



0166_H_PULMFON_SVD

Legacy Name: PT3_6M

Model added: 31 Jan 2023

Species	Human
Anatomy	Pulmonary Fontan
Disease	Single Ventricle Defect
Procedure	Fontan TEVG

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.

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, is not 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 the intact ventricular septum (PA/IVS), and atrioventricular canal defect (AV Canal).

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.

TEVG

TEVGs, or Tissue Engineered Vascular Grafts, are becoming a more popular alternative to traditional grafts made of synthetic materials. The general approach to constructing a TEVG involves taking human-harvested cells and then seeding them into a scaffold. These TEVGs can then be used in various operations such as the Fontan or Glenn procedures and have the advantage over synthetic grafts of being able to grow and remodel over time with the host body.

Clinical Data

General Patient Data

Age (yrs)	2.5
Sex	Female

Specific Patient Data

Patient diagnosis: HLHS, mitral atresia/aortic atresia variant.

Graft diameter at implant: 16mm

Elastic moduli were tuned to match fraction area change deformation observed at each MRI imaging session.

Ethnicity	Caucasian
Time post Fontan operation (months)	6

Minimum graft diameter (mm)	7.1
Diameter stenosis (% from implant)	56
Area stenosis (% from implant)	79
BSA (m^2)	0.53
Elastic modulus (kPa)	16

Notes

Model of a patient's pulmonary arteries 6 months after they underwent a modified Fontan procedure where a tissue-engineered vascular graft was used to connect the inferior vena cava (IVC) to the pulmonary arteries. Patient number 3 in the study. \nSee paper for more details. See below for information on the image data.

Image Modality: MR

Image Type: VTI

Image Source: Nationwide Children's Hospital, Columbus, OH

Publications

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

Schwarz, E.L., Kelly, J.M., Blum, K.M. et al. Hemodynamic performance of tissue-engineered vascular grafts in Fontan patients. npj Regen Med 6, 38 (2021). https://doi.org/10.1038/s41536-021-00148-w

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