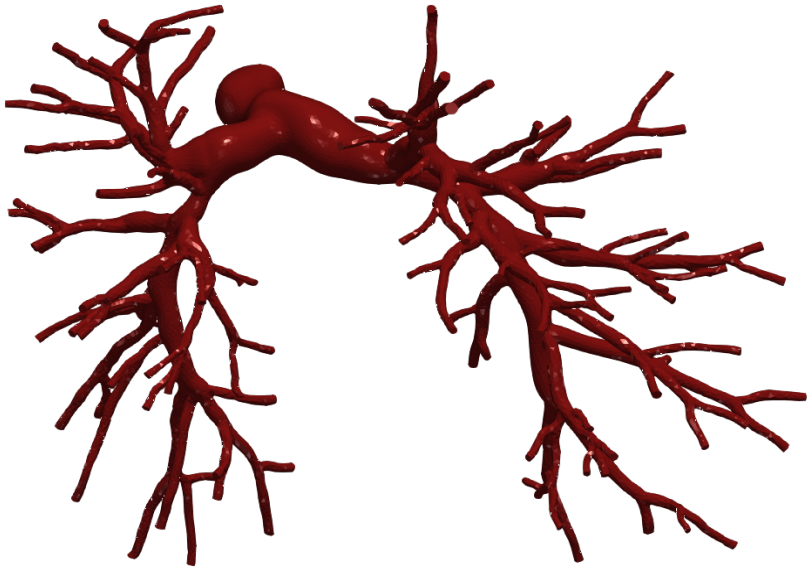
Vascular Model Repository Specifications Document



0119_0001

Species	Human
Anatomy	Pulmonary
Disease	Alagille Syndrome
Procedure	-

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.

Alagille Syndrome

Alagille syndrome (ALGS) is a rare genetic disorder that can affect multiple organ systems of the body including the liver, heart, skeleton, eyes and kidneys. The specific symptoms and severity of Alagille syndrome can vary greatly from one person to another, even within the same family. Common symptoms, which often develop during the first three months of life, include blockage of the flow of bile from the liver (cholestasis), yellowing of the skin and mucous membranes (jaundice), poor weight gain and growth, and severe itching (pruritis). Additional symptoms include heart murmurs, congenital heart defects, vertebral (back bone) differences, thickening of the ring that normally lines the cornea in the eye (posterior embryotoxon) and distinctive facial features. The current estimated incidence of ALGS is approximately 1/30,000 to 1/45,000.

Many individuals with Alagille syndrome have heart (cardiac) abnormalities that can range from benign heart murmurs to serious structural defects. A heart murmur is an extra sound that is heard during a heartbeat. Heart murmurs in children with Alagille syndrome are usually caused by narrowing of the blood vessels of the lungs (pulmonary artery stenosis). The most common heart abnormality is peripheral pulmonary stenosis in which some of the blood vessels carrying blood to the lungs (pulmonary arteries) are narrowed (stenosis). Some children with Alagille syndrome

may have complex heart defects, the most common of which is tetralogy of Fallot.

Clinical Data

General Patient Data

Age (yrs)	16
Sex	Male

Specific Patient Data

BSA (m^2)	1.2
Height (m)	1.49
Weight (kg)	31.4
CI (L/min/m^2)	2.77
P LPA MP cath	15
P LPA SP cath	30
P LPA DP cath	6
P RPA MP cath	14
P RPA SP cath	27
P RPA DP cath	6
F PA cath	4.62

Notes

- See below for information on the image data and boundary conditions associated with the model.

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Publications

There are no publications associated with the featured model.

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