

# Vascular Model Repository

## Specifications Document



0119\_0001

<b>Species</b>	Human
<b>Anatomy</b>	Pulmonary
<b>Disease</b>	Alagille Syndrome
<b>Procedure</b>	-

# 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 <sup>2</sup> )	1.2
Height (m)	1.49
Weight (kg)	31.4
CI (L/min/m <sup>2</sup> )	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.

**Image Modality:** CT

**Image Type:** DICOM

**Image Source:** STAN

**Image Manufacturer:** SIEMENS

## Publications

There are no publications associated with the featured model.

# License

Copyright (c) Stanford University, the Regents of the University of California, Open Source Medical Software Corporation, and other parties.

All Rights Reserved.

Permission is hereby granted, free of charge, to any person obtaining a copy of this data to use the data for research and development purposes subject to the following conditions:

The above copyright notice and the README-COPYRIGHT file shall be included in all copies of any portion of this data. Whenever reasonable and possible in publications and presentations when this data is used in whole or part, please include an acknowledgement similar to the following:

"The data used herein was provided in whole or in part with Federal funds from the National Library of Medicine under Grant No. R01LM013120, and the National Heart, Lung, and Blood Institute, National Institutes of Health, Department of Health and Human Services, under Contract No. HHSN268201100035C"

AND/OR

N.M. Wilson, A.K. Ortiz, and A.B. Johnson, "The Vascular Model Repository: A Public Resource of Medical Imaging Data and Blood Flow Simulation Results," J. Med. Devices 7(4), 040923 (Dec 05, 2013) doi:10.1115/1.4025983.

AND/OR

Reference the official website for this data: [www.vascularmodel.com](http://www.vascularmodel.com)

THE DATA IS PROVIDED "AS IS", WITHOUT WARRANTY OF ANY KIND, EXPRESS OR IMPLIED, INCLUDING BUT NOT LIMITED TO THE WARRANTIES OF MERCHANTABILITY, FITNESS FOR A PARTICULAR PURPOSE AND NONINFRINGEMENT. IN NO EVENT SHALL THE AUTHORS OR COPYRIGHT HOLDERS BE LIABLE FOR ANY CLAIM, DAMAGES OR OTHER LIABILITY, WHETHER IN AN ACTION OF CONTRACT, TORT OR OTHERWISE, ARISING FROM, OUT OF OR IN CONNECTION WITH THE DATA OR THE USE OR OTHER DEALINGS IN THE DATA.