

Vascular Model Repository

Specifications Document



0119_0001

Species	Human
Anatomy	Pulmonary
Disease	Alagille Syndrome
Procedure	-

Clinical Significance and Background

Pulmonary

The pulmonary arteries are blood vessels that carry systemic venous blood returning to the right side of the heart through to the microcirculation of the lungs. Unlike in other organs where arteries supply oxygenated blood, the blood carried by the pulmonary arteries is deoxygenated, as it is venous blood returning to the heart. The main pulmonary arteries emerge from the right side of the heart, and then split into smaller arteries that progressively divide and become arterioles, eventually narrowing into the capillary microcirculation of the lungs where gas exchange occurs.

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.

Image Modality: CT

Image Type: DICOM

Image Source: STAN

Image Manufacturer: SIEMENS

Publications

There are no publications associated with the featured model.

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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

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