

Vascular Model Repository

Specifications Document



0116_H_PULM_ALGS

Legacy Name: AS3_SU0238_prestent

Model added: 3 Aug 2022

Species	Human
Anatomy	Pulmonary
Disease	Alagille Syndrome
Procedure	None

Clinical Significance and Background

Pulmonary

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 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 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 (backbone) 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 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 the 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 are narrowed. 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)	13.2
Sex	Male

Specific Patient Data

Heart Rate (bpm)	78
BSA (m ²)	1
Cardiac Index (L/min/m ²)	2.6
Wedge Pressure (mmHg)	17
MPA pressures: systolic/diastolic (mmHg)	68/15
MPA mean pressure (mmHg)	38
RPA pressures: systolic/diastolic (mmHg)	39/12
RPA mean pressure (mmHg)	22
LPA pressures: systolic/diastolic (mmHg)	29/15
LPA mean pressure (mmHg)	21
PA flow split, percentage right	68%

Notes

Model of a patient suffering from peripheral pulmonary stenosis from Alagille syndrome. This model is before any in-silico stenting procedures have been done. \n\nThe corresponding model with in-silico generated stents can be found in the VMR as 0122_H_PULM_ALGS. \n\nSee [paper](#) for more details. See below for information on the image data.

Image Modality: CT/MR

Image Type: VTI

Image Source: Lucille Packard Children's Hospital

Publications

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

Lan, I. S., Yang, W., Feinstein, J. A., Kreutzer, J., Collins, R. T., Ma, M., ... & Marsden, A. L. (2022). Virtual Transcatheter Interventions for Peripheral Pulmonary Artery Stenosis in Williams and Alagille Syndromes. *Journal of the American Heart Association*, 11(6), e023532.

<http://www.doi.org/10.1161/JAHA.121.023532>

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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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