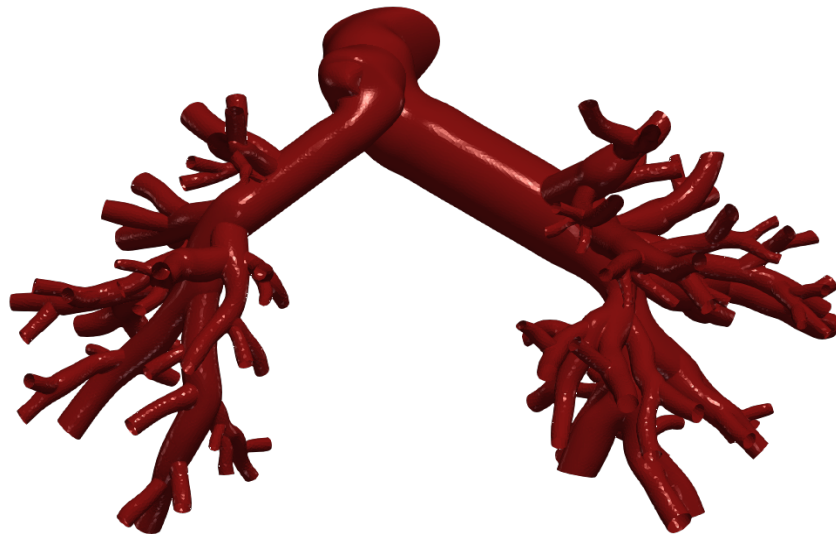


# Vascular Model Repository

## Specifications Document



### 0123\_H\_PULM\_ALGS

Legacy Name: AS4\_SU0287\_stent

Model added: 23 Aug 2022

|                  |                   |
|------------------|-------------------|
| <b>Species</b>   | Human             |
| <b>Anatomy</b>   | Pulmonary         |
| <b>Disease</b>   | Alagille Syndrome |
| <b>Procedure</b> | 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) | 5.8  |
| Sex       | Male |

## Notes

Model of a patient suffering from peripheral pulmonary stenosis from Alagille syndrome. \nNOTE: These models are based on artificially generated stents that are used to simulate the effectiveness of proximal and extensive stenting in combatting PPS in Alagille syndrome. The image files are still based on the original diseased patient, but the model and mesh files have been manually edited where proximal stenting and extensive stenting models have been created and are in separate folders. \nThe corresponding original model without the generated stent is 0117\_H\_PULM\_ALGS. \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](http://www.vascularmodel.com)

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