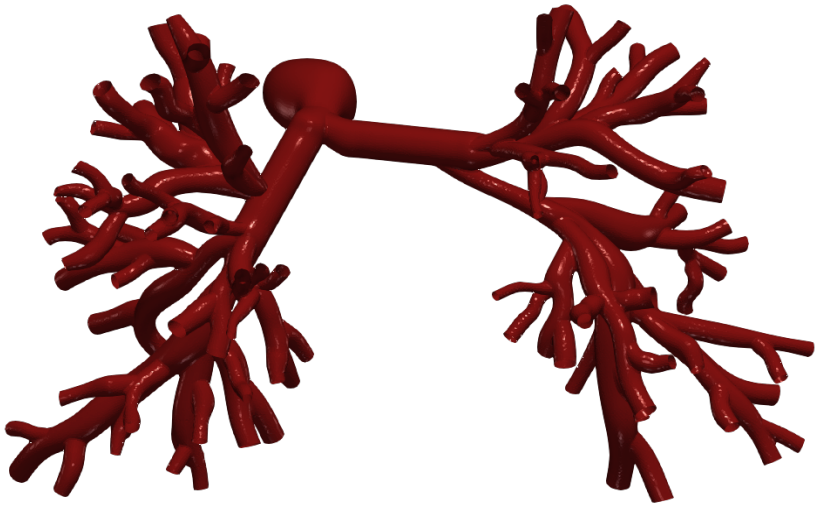
Vascular Model Repository Specifications Document



0124_H_PULM_WS

Legacy Name: WS1_SU0267_stent

Model added: 23 Aug 2022

Species	Human
Anatomy	Pulmonary
Disease	Williams 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.

Williams Syndrome

Williams syndrome, also known as Williams-Beuren syndrome, is a rare genetic disorder characterized by growth delays before and after birth (prenatal and postnatal growth retardation), short stature, a varying degree of mental deficiency, and distinctive facial features that typically become more pronounced with age. Newborns with Williams syndrome have characteristic elfin-like facial features including an unusually small head (microcephaly), full cheeks, an abnormally broad forehead, puffiness around the eyes and lips, a depressed nasal bridge, broad nose, and/or an unusually wide and prominent open mouth.

Congenital heart defects (CHD) occur in approximately 75 percent of children with Williams syndrome. The most frequent defect is supravalvar aortic stenosis, a condition characterized by the narrowing of the aorta above the aortic valve. Additional congenital heart defects associated with Williams syndrome may include pulmonary artery stenosis, and/or septal defects. Abnormally high blood pressure (hypertension) is also common in adults with this disorder.

Clinical Data

General Patient Data

Age (yrs)	0.63
Sex	Male

Notes

Model of a patient suffering from peripheral pulmonary stenosis from Williams 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 Williams 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 0118_H_PULM_WS. \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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