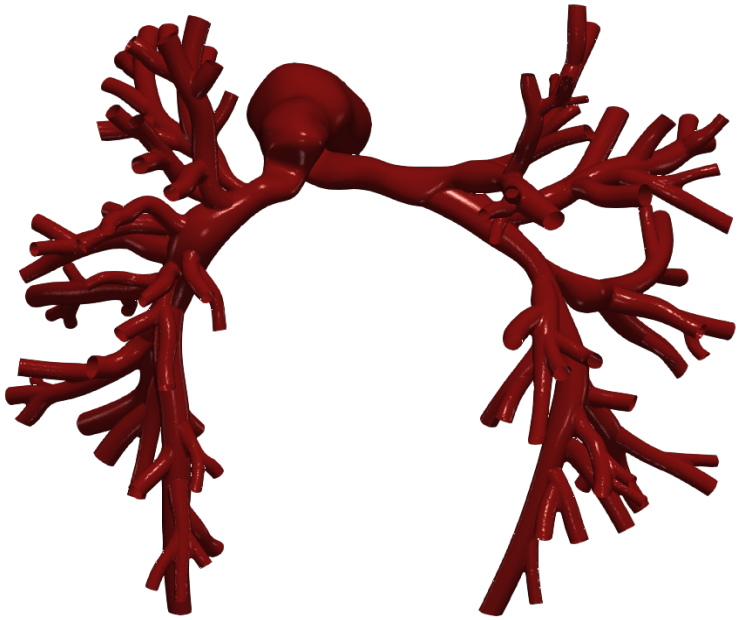
Vascular Model Repository Specifications Document



0119_H_PULM_WS

Legacy Name: WS2_SU0269_prestent

Model added: 3 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)	0.2
Sex	Female

Specific Patient Data

Heart Rate (bpm)	122
BSA (m^2)	0.25
Cardiac Index (L/min/m^2)	4.08
Wedge Pressure (mmHg)	12
MPA pressures: systolic/diastolic (mmHg)	93/42
MPA mean pressure (mmHg)	54
RPA pressures: systolic/diastolic (mmHg)	20/10
RPA mean pressure (mmHg)	17
LPA pressures: systolic/diastolic (mmHg)	27/16
LPA mean pressure (mmHg)	20

Notes

Model of a patient suffering from peripheral pulmonary stenosis from Williams syndrome. This model is before any in-silico stenting procedures have been done. \nThe corresponding model with in-silico generated stents can be found in the VMR as 0125_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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