Unrepaired persistent truncus arteriosus in an adult

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History

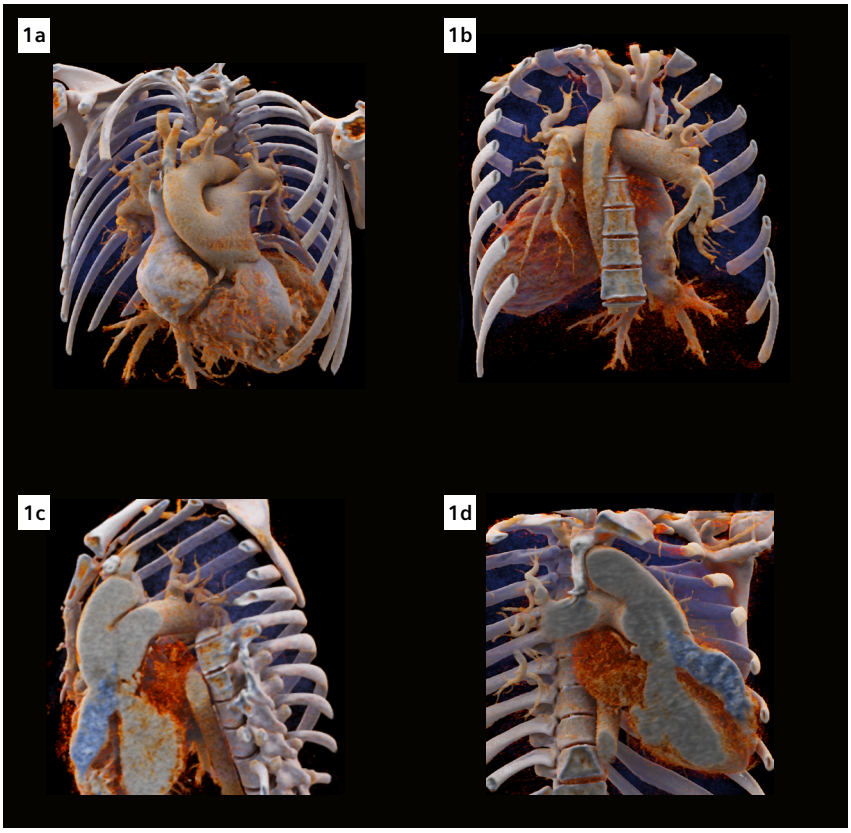
A 27-year-old female patient came to the hospital complaining of a mild edema developing in both of her legs, for no obvious reason, over the past two weeks. Otherwise, she reported coughing with white phlegm, occasional dizziness and headaches as well as exertional dyspnea that could be relieved by rest. The physical examination revealed both central and peripheral cyanosis. A previous diagnosis of "congenital heart disease" in her childhood was reported, however no details could be provided. A chest CT with contrast was requested for further evaluation.

Diagnosis

CT images revealed a common arterial trunk, overriding a high ventricular septal defect (VSD), shared by the ascending aorta (AA) and the main pulmonary artery (MPA), compatible with truncus arteriosus (TA) Collett & Edwards type I or Van Praagh type A1. The MPA, originating from the left lateral aspect of the common trunk, divided into the right and the left pulmonary arteries (PA). Signs of central PA dilatation, tapering of peripheral PA, cardiomegaly with biventricular hypertrophy and marked enlargement of the right atrium and left ventricle, along with a mosaic pattern of attenuation in the lungs, consistent with severe chronic pulmonary hypertension, were visualized. The aortic arch and the brachiocephalic arteries showed no abnormalities. The left coronary artery (LCA) originated off the right-posterior aspect of the TA, coursing between the left atrium and the TA. The origin and the course of the right coronary artery (RCA) appeared normal.

Subsequently, an echocardiography examination confirmed the CT findings and additionally revealed a mild regurgitation of all four valves.

The patient was only treated medically, due to her pulmonary condition, and was discharged with noticeable improvement one week later.



1 cVRT images show a common arterial trunk, overriding a high VSD, shared by the AA and the MPA. The MPA, originating off the left lateral aspect of the common trunk, divides into the right and the left PA. Dilatation of the central PA and tapering of peripheral PA are visualized, consistent with severe chronical pulmonary hypertension. Cardiomegaly with marked right atrium and left ventricle enlargement is also seen. The aortic arch and the brachiocephalic arteries are normal.

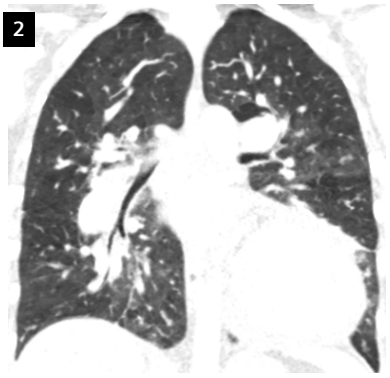
Comments

Persistent truncus arteriosus (PTA), which is often referred to simply as TA, is a rare congenital heart disease. A TA is an embryological structure which should properly divide itself into the aorta and the pulmonary trunk. Failure to do so results in only one arterial trunk arising from the heart, usually overriding a VSD, providing mixed oxygenated and carbonized blood to both the aortic and pulmonary circulation. Early surgical correction is the treatment of choice and can guarantee long-term survival since over 80% of infants with TA die within their first life year. [1] [2]
In rare cases, a person with TA survives infancy without surgical repair and lives into adulthood, such as the present case. However, these patients have a high risk for pulmonary hypertension and the Eisenmenger syndrome. Echocardiography is the primary imaging examination to comprehend dynamic blood flow and valve function; whereas the strength of CT lies in the ability to describe anatomical details, such as the origin and the course of the coronary arteries and to determine the exact course and relation of the greater arteries to each other, both mandatory for exact presurgical planning. In the present case, the use of only 70 kV tube voltage was feasible due to the patient's body habitus. This lower kV setting has the advantage of an increased contrast-to-noise ratio in CT angiography with a lower radiation dose compared to a 120 kV protocol. Furthermore, the amount of required contrast material can be significantly reduced (30 mL). Cinematic volume rendering allows for a realistic 3D display of the anatomy with improved depth and shape perceptions. This enables an optimal overview of the anatomical structures which is important for treatment planning, communication with the patient and between physicians. ●

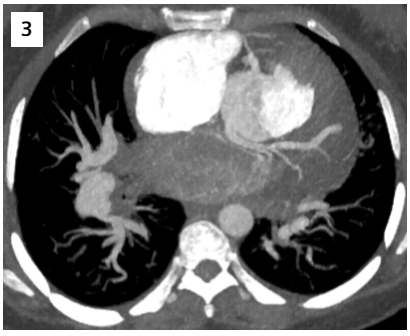
References

[1] F. Guenther, et al. Persistent truncus arteriosus: a rare finding in adults. European Heart Journal, Volume 30, Issue 9, May 2009, Page 1154. doi.org/10.1093/eurheartj/ehp020.

[2] Phillip S. Naimo, Igor E. Konstantinov. Surgery for Truncus Arteriosus: Contemporary Practice. Ann Thorac Surg 2021;111:1442-50.



2 **A coronal MPR image shows a mosaic pattern of attenuation in the lungs and cardiomegaly.**



3 **An axial MIP image shows a normal origin of the RCA, and a LCA originating off the right-posterior aspect of the TA, coursing between the left atrium and the TA.**

Examination Protocol

Scanner	**SOMATOM Force**
Scan area	Thorax
Scan mode	Spiral mode
Scan length	182 mm
Scan direction	Cranio-caudal
Scan time	1 s
Tube voltage	70 kV
Effective mAs	169 mAs
Dose modulation	CARE Dose4D
$CTDI_{vol}$	1.98 mGy
DLP	67.1 mGy*cm

Rotation time	0.25 s
Pitch	1.2
Slice collimation	192 x 0.6 mm
Slice width	0.6 mm
Reconstruction increment	0.5 mm
Reconstruction kernel	Bv36, ADMIRE 3
Contrast	**350 mg/mL**
Volume	30 mL + 40 mL saline
Flow rate	3.5 mL/s
Start delay	Bolus tracking triggered at 70 HU in the MPA + 3 s

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