"Crossed" pulmonary arteries in a newborn with truncus arteriosus: An unusual anatomic variant

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ABSTRACT

We report a newborn with truncus arteriosus and a very unusual variant of "crossed" pulmonary arteries. The left pulmonary artery arose anterior and slightly to the right side of the common trunk and the right pulmonary artery arose from the posterior and left side of the common trunk. Computed tomographic images and a diagram are provided.

Keywords: Congenital heart disease, crossed pulmonary arteries, truncus arteriosus

CLINICAL SUMMARY

A fetal echocardiogram performed at 26 weeks was suggestive of truncus arteriosus. The newborn echocardiogram showed a dysplastic truncal valve and a right-sided aortic arch. The right pulmonary artery originated posteriorly and to the left of the truncus. The left pulmonary artery originated anteriorly and slightly to the right of the truncus. A computed tomographic scan confirmed the findings [Figure 1]. The patient underwent surgical repair at 10 days of age, with right pulmonary artery unifocalization to the left pulmonary artery. The left pulmonary artery was left attached to the truncus. The right pulmonary was resected and excised from the truncus. An arteriotomy was made in the distal left pulmonary artery, just proximal to the hilar branches. The right pulmonary artery was attached to the left pulmonary arteriotomy site. The proximal left pulmonary artery was incised inferiorly. The aortopulmonary window was patch closed. The distal right ventricle to pulmonary artery homograft was attached to the underside of a patch-augmented left pulmonary artery. In the next 6 weeks, the truncal valve regurgitation progressed from mild to severe, and because of persistent significant bilateral pulmonary artery stenoses (gradient >50 mmHg), she underwent reoperation at 2 months of age with pulmonary artery plasty, homograft replacement, and truncal valve replacement with a15 mm St. Jude valve. At 6 months of age, an echocardiogram showed residual bilateral pulmonary artery stenoses, with a peak velocity of 3.5 m/s on the right and 2.7 m/s on the left. The plan is for a cardiac catheterization at a year of age with possible intervention on the branch pulmonary arteries.

DIFFERENTIAL DIAGNOSIS

At the time of birth, the differential diagnosis also included hemitruncus and tetralogy of Fallot with a right aortic arch and multiple aortopulmonary collaterals. The initial echocardiogram confirmed the diagnosis. The more important dilemma in this patient was how to incorporate the pulmonary arteries during the first surgical intervention.

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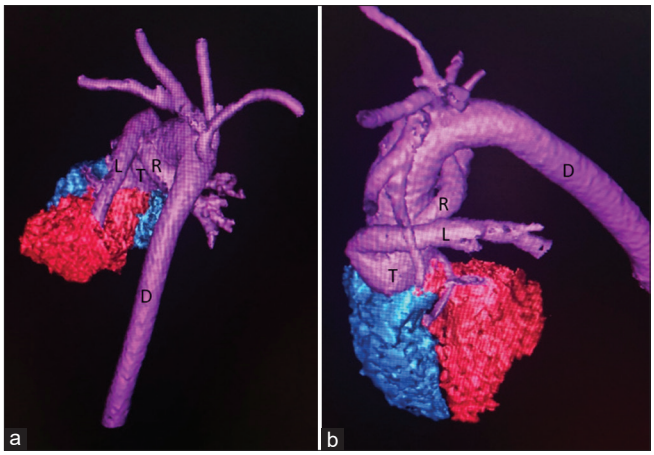

Figure 1: Computed tomographic reconstructed images. (a) Posterior view of the heart. (b) Left lateral view. T: Truncus arteriosus, L: Left pulmonary artery, R: Right pulmonary artery, D: Descending aorta

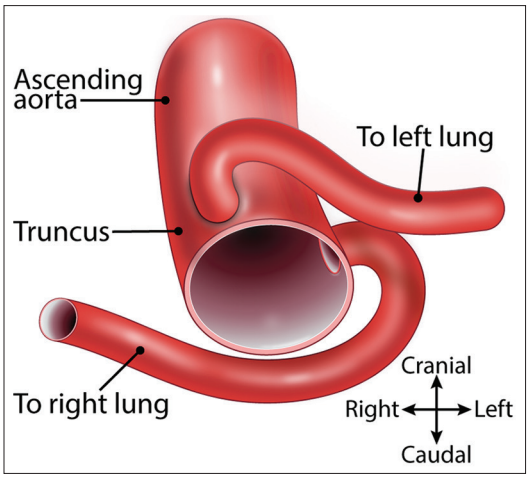

Figure 2: Diagramatic representation of the crossed pulmonary arterial anatomy, caudal, and anterior view

DISCUSSION

The pulmonary arteries in truncus arteriosus usually originate from a main pulmonary artery or from the truncus arteriosus ipsilaterally.[1] Crossed pulmonary arteries are very rare. Butto *et al.* described crossed pulmonary arteries in 3 of 54 pathologic truncus arteriosus cases.[2] Crossed pulmonary arteries were first described by Jue *et al.* in 1996 in a patient with trisomy 18, an atrial septal defect and anomalous pulmonary venous return.[3] The authors speculated that the anatomy could have been caused by faulty differential growth within the pulmonary trunk and counterclockwise rotation of the normal origins of the pulmonary arteries. Our patient's anatomy appears to be consistent with abnormal counterclockwise rotation of about 120° of the common trunk [Figure 2]. "Crossed" or "malposed" pulmonary arteries have been described more frequently in patients with DiGeorge syndrome, deletions of chromosomal region 22q11,[4] a right aortic arch, or an interrupted aortic arch.[5] Crossed pulmonary arteries constitute a challenge to surgical repair. Creative and aggressive pulmonary artery mobilization, unifocalization, and plasty may be necessary to avoid the need for repeat intervention.

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Conflicts of interest

There are no conflicts of interest.

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