Radiología. 2017;59(3):209–217

RADIOLOGÍA

www.elsevier.es/rx

RADIOLOGY THROUGH IMAGES

Congenital anomalies of the pulmonary arteries: Spectrum of findings on computed tomography☆

J. Bueno[a,*], **L. Flors**[a], **M. Mejía**[b]

[a] *Imagen Cardiotorácica, Departamento de Radiología e Imagen Médica, Universidad de Virginia, Charlottesville, VA, United States*
[b] *Departamento de Radiología, Fundación Valle del Lili, Cali, Valle del Cauca, Colombia*

Received 25 April 2016; accepted 16 September 2016

KEYWORDS
Pulmonary arteries;
Congenital anomalies;
Pulmonary agenesis;
Anomalous origin of the pulmonary artery;
Tetralogy of Fallot;
Computed tomography

Abstract
Objective: Congenital anomalies of the pulmonary arteries are uncommon. They can occur in isolation or in association with congenital heart defects. Isolated congenital anomalies remain undiscovered until they are reported as incidental findings on imaging tests, usually not until adolescence. We review the embryological development and normal anatomy of the pulmonary arteries as well as the spectrum of computed tomography findings for various congenital anomalies: unilateral interruption of the pulmonary artery, anomalous origin of the left pulmonary artery (pulmonary artery sling), idiopathic aneurysm of the pulmonary artery, and other anomalies associated with congenital heart defects.
Conclusion: Congenital anomalies of the pulmonary arteries represent a diagnostic challenge for clinicians and radiologists. Computed tomography is useful for their diagnosis, and general radiologists need to be familiar with their imaging appearance because they are often discovered incidentally.

PALABRAS CLAVE
Arterias pulmonares;
Anomalías congénitas;

Anomalías congénitas de las arterias pulmonares: espectro de hallazgos en tomografía computarizada

Resumen Las anomalías congénitas de las arterias pulmonares (AP) son poco frecuentes y pueden presentarse aisladas o asociadas a defectos cardiacos congénitos. En general, si son

☆ Please cite this article as: Bueno J, Flors L, Mejía M. Anomalías congénitas de las arterias pulmonares: espectro de hallazgos en tomografía computarizada. Radiología. 2017;59:209–217.

* Corresponding author.
E-mail address: julianabueno.m@gmail.com (J. Bueno).

Agenesia pulmonar;
Origen anómalo de la arteria pulmonar;
Tetralogía de Fallot;
Tomografía computarizada

aisladas, suelen pasar clínicamente inadvertidas hasta la adolescencia y ser un hallazgo incidental en exploraciones radiológicas. Nuestro objetivo es revisar las bases embriológicas del desarrollo de las AP, su anatomía normal y el espectro de hallazgos por tomografía computarizada (TC) de sus anomalías congénitas, en concreto la interrupción unilateral de la AP, el origen anómalo de la AP izquierda (*sling* pulmonar), el aneurisma idiopático de la AP y otras anomalías asociadas a defectos cardiacos congénitos. Las anomalías congénitas de las AP representan un reto diagnóstico, tanto clínico como radiológico. La TC es una herramienta útil en su diagnóstico y el radiólogo general debe estar familiarizado con su apariencia, ya que pueden ser un hallazgo incidental.

Introduction

Congenital anomalies of pulmonary arteries (PA) are usually associated to other congenital heart defects and they do not usually occur in isolation. Although they can result in symptoms during childhood, the PA isolated anomalies usually go unnoticed until adolescence or early adult age.[1,2]

The imaging diagnosis of PA anomalies are challenging both for the clinician and the radiologist since conventional image modalities like X-rays and ultrasounds have a limited value when it comes to detection and characterization, while the pulmonary angiography is an invasive method usually reserved for cases that require treatment. This is the reason why both the computed tomography (CT) and magnetic resonance imaging (MRI) play an essential role in the study of PA. In part thanks to the possibility of generating particular multiplanar and volumetric reconstructions the angio-CT offers great advantages in the diagnosis and follow-up of these anomalies making the characterization of the vascular and valvular anatomies possible as well as any other associated findings in other organs. Also these reconstructions facilitate the communication of findings and the understanding of the anatomy for an optimal surgical plan.

The goal of this paper is to review the embyological basis for the development of PA, the PA normal anatomy and image findings of PA congenital anomalies—especially the unilateral interruption of PA, the anomalous origin of the left PA (pulmonary sling), the main PA idiopathic aneurysm and other anomalies associated with congenital heart anomalies.

Embryological basis

After the formation of the cardiac tube and its differentiation into primitive segments that give rise to cardiac structures the arterial trunk gives rise to the aorta and the proximal section of the PA.[3] During the fifth week of embryonic life the aortopulmonary septum is created in a spiral-like shape that splits the truncus arteriosus into aorta and PAs (Fig. 1).

Thus the trunk of the pulmonary artery is derived into the truncus arteriosus. The proximal sections of the sixth bilateral aortic arches make up the right and left Pas.[4] The distal sections of the PAs are derived from primitive buds

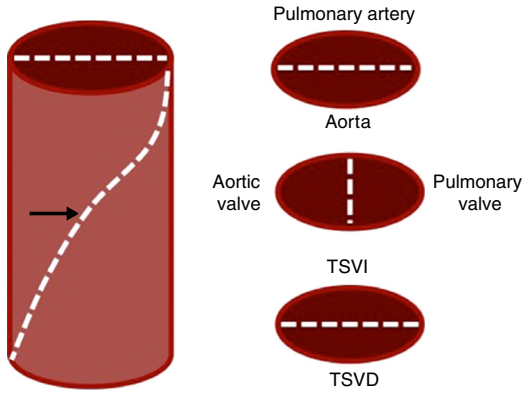

Figure 1 Embryonic development of pulmonary artery. Schematic illustration of the arterial trunk (left). The aortopulmonary septum (arrow) shows a spiral disposition and divides the arterial trunk into aorta and pulmonary aorta as well as the aortic and pulmonary valves while dividing the outflow track of both ventricles inferiorly. RVOT: right ventricle outflow track; LVOT: left ventricle outflow track.

that come from these arches and grow toward the inside of lung buds (the origin of lungs) to later anastomose with them.

We should remember that: since the embryonic lung morphogenesis is a process parallel to the airway branching morphogenesis,[4] it is not rare to find a combination of vascular and bronchial tree anomalies.

The anatomy of pulmonary arteries

The PA trunk stems from the right ventricle and bifurcates into the main right and left PAs. The pulmonary valve is made up of three valvular sinuses and can be found in the proximal section of the pulmonary trunk—the pulmonary root.[5]

The maximum diameter of the adult pulmonary trunk is 28 mm.[6] The right and left PAs show an intrapericardial trajectory before bifurcating into the lobar arteries in every side. Because of the special location of the pulmonary trunk—left to the middle line, the intrapericardial segment of the right PA is longer ant its bifurcation occurs at pulmonary root level. The right PA is located under the right