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Author for correspondence:
E. Martínez-Quintana, MD, PhD, Servicio de Cardiología, Complejo Hospitalario Universitario Insular Materno Infantil, Avd. Marítima del Sur s/n., 35016 Las Palmas de Gran Canaria, Spain. Tel: 0034928441360;
E-mail: efrencardio@gmail.com

Giant right pulmonary artery aneurysm in a systemic-to-pulmonary artery shunt

Efrén Martínez-Quintana[1,2], Jose M. Medina-Gil[1] and Jorge Marrero-Brito[3]

[1]Cardiology Service, Insular-Materno Infantil University Hospital, Las Palmas de Gran Canaria, Spain; [2]Medical and Surgical Sciences Department, Faculty of Health Sciences, University of Las Palmas de Gran Canaria, Las Palmas de Gran Canaria, Spain and [3]Radiology Service, Insular-Materno Infantil University Hospital, Las Palmas de Gran Canaria, Spain

Abstract

Aneurysms of the pulmonary arteries and trunk are rare entities. The Waterston shunt is a palliative procedure for children with cyanotic CHD due to obstruction of the pulmonary outflow. Described complications are distortion of the pulmonary artery and pulmonary arterial hypertension. We report a patient with a giant right pulmonary artery aneurysm in relation to a Waterston shunt.

A 39-year-old male patient with a pulmonary atresia who underwent construction of a Waterston shunt as neonate was referred to our CHD unit for clinical evaluation. He had an NYHA functional class III with associated cyanosis (basal oxygen saturation of 64%) and a pulmonary artery aneurysm with non-specific symptoms. A previous chest x-ray, carried out 13 years before, showed a right pulmonary artery aneurysm (Fig 1a) that had increased in size to 72 × 100 × 136 mm as it may be seen in the current chest x-ray (Fig 1b) and thoracic CT (Fig 1c). Furthermore, pulmonary catheterisation evidenced permeability of the shunt and a giant pulmonary artery aneurysm located at the right pulmonary artery (Fig 1d). Haemodynamic data showed a right pulmonary artery pressure of 40/30 (34) mmHg. Although the patient was "clinically stable" and we opted for a conservative management due to his high surgical risk he presented a *progressive clinical deterioration* dying as a result of heart failure, multi-lobar pneumonia, and multi-resistant pseudomonas aeruginosa infection during a hospital admission. However, his right pulmonary artery aneurysm was not deemed a factor in the patient's demise.

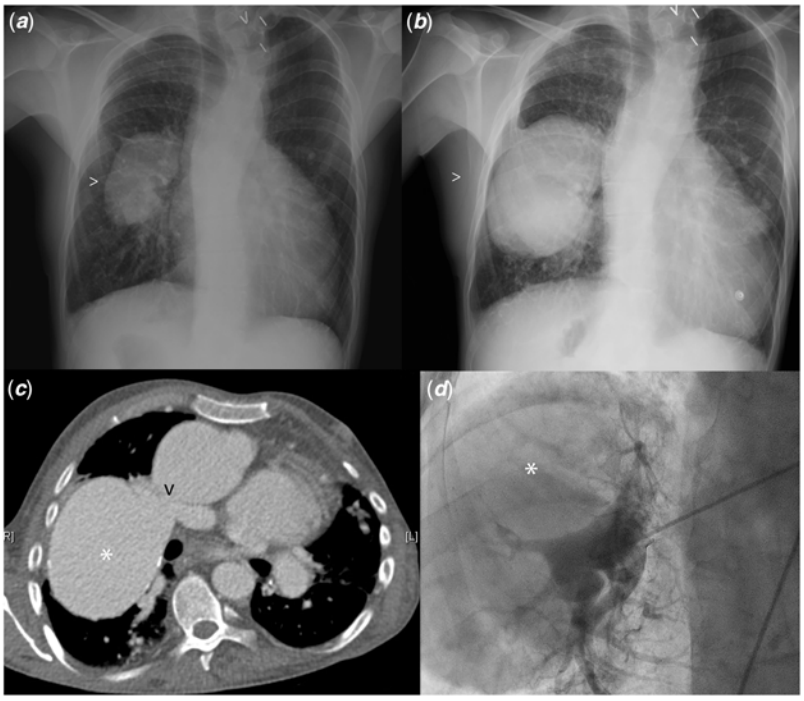

Figure 1. Radiological findings of the giant right pulmonary artery aneurysm. Arrow heads in figures (***a***) and (***b***) and asterisk in figures (***c***) and (***d***) point to the right pulmary artery aneurysm. The arrow head in (***c***) shows the connection of the Waterston shunt to the right pulmonary artery and the pulmonary artery aneurysm.

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Aneurysms of the pulmonary arteries and trunk are rare entities. They have been associated with CHD, structural vascular anomalies, vasculitis, arterial pulmonary hypertension, penetrating and blunt trauma, or infection.[1] Anastomosis of the ascending aorta to the right pulmonary artery, the so-called Waterston shunt, has been undertaken as a palliative procedure for children with cyanotic CHD due to obstruction of the pulmonary outflow tract with reduced pulmonary blood flow. Haemodynamic complications may occur because of shunt of excessive size leading initially to excessive pulmonary blood flow, left ventricular overwork, and congestive heart failure and subsequently to pulmonary arterial hypertension.[2] Over time, this pulsatile and turbulent flow through the shunt may also promote and raise shear stress on the adjacent pulmonary artery with the consequent weakness and development of pulmonary aneurysms,[3,4] pulmonary arterial dissections,[5] false aneurysms[6] or kinking, and distortion of the pulmonary artery besides technical difficulties with takedown. Because pulmonary artery aneurysms may lead to patient's death[7,8] some authors[9] recommend surgical repair if the aneurysms are large (>5.5 cm) or if they are symptomatic, regardless of the size, because of the risk of rupture or dissection according to the guidelines for aortic disease. However, this cut-off value has its limits as the literature tends to reflect a positive reporting bias and there is no clear guideline for the best therapeutic approach due to the infrequency of the disease.

Author ORCIDs. Efrén Martínez-Quintana, 0000-0002-1289-7279

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Conflicts of Interest. None.

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