Pulmonary sling in a patient with common arterial trunk

Mohamed Sobh[1], Patrick Langguth[2], Kolja Becker[1], Jens Scheewe[3], Anselm Uebing[1], Inga Voges[1]

[1]Department of Congenital Heart Disease and Paediatric Cardiology, University Hospital Schleswig-Holstein, Kiel, Germany, [2]Department of Radiology and Neuroradiology, University Hospital Schleswig-Holstein, Kiel, Germany, [3]Department of Cardiovascular Surgery, University Hospital Schleswig-Holstein, Kiel, Germany

ABSTRACT

We report a rare association of common arterial trunk with left pulmonary artery sling and highlight the importance of cross-sectional imaging in complex congenital cardiac lesions. The patient was antenatally diagnosed with common arterial trunk and underwent surgical repair in the neonatal period. At the age of 20 months, the patient presented with respiratory symptoms and increased right ventricular pressure. Multislice computed tomography demonstrated a pulmonary sling with compression of the distal trachea. Surgical correction of the pulmonary sling and change of the right ventricular to pulmonary artery conduit to a bigger size was performed.

Keywords: Common arterial trunk, computed tomography, cross-sectional imaging, pulmonary sling

INTRODUCTION

Pulmonary artery sling is a rare congenital vascular anomaly that is often associated with complete tracheal rings. The left pulmonary artery originates from the posterior aspect of the right pulmonary artery and then passes between the trachea and esophagus toward the left lung.[1] Approximately 90% of patients with left pulmonary artery sling are symptomatic within the 1st year of life. Symptoms include signs of extrinsic airway obstruction such as stridor and dyspnea and recurrent pneumonia.[2] Pulmonary sling may be associated with congenital heart disease, however only few reports about patients with common arterial trunk and pulmonary sling exist.[3-5]

CLINICAL SUMMARY

We report the case of a 20-month-old girl who was diagnosed antenatally with common arterial trunk and a balanced division of the trunk into the systemic and pulmonary pathways, which was confirmed after birth [Figure 1]. Surgery was performed in the neonatal period with excision of the pulmonary artery segment from the common trunk followed by mobilization of the pulmonary arteries behind the aorta and implantation of an 8-mm conduit from the right ventricle to the pulmonary arteries. It was noted that the pulmonary artery segment was long, that there was a large persistent left superior vena cava and that the pulmonary bifurcation was far behind the aorta. There was no sinusal origin of the pulmonary artery segment. The distal right and left pulmonary arteries were not visualized and not followed deep into the chest. The ventricular septal defect was closed with a patch.

From the age of 10 months, there was a steady decline in physical activity, and the patient presented with stridor and exertional dyspnea. In addition, she experienced

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Quick Response Code:

Website: www.annalspc.com

DOI: 10.4103/apc.APC_98_20

How to cite this article: Sobh M, Langguth P, Becker K, Scheewe J, Uebing A, Voges I. Pulmonary sling in a patient with common arterial trunk. Ann Pediatr Card 2021;14:239-41.

Address for correspondence: Dr. Inga Voges, Department of Congenital Heart Disease and Paediatric Cardiology, Arnold-Heller-Street 3, University Hospital Schleswig-Holstein, Campus Kiel, Kiel, Germany.
E-mail: inga.voges@uksh.de

Submitted: 04-May-2020 **Revised:** 20-Jun-2020 **Accepted:** 20-Oct-2020 **Published:** 30-Dec-2020

frequent respiratory infections including severe mycoplasma pneumonia. External ear-nose-throat exam did not provide sufficient explanation for the stridor. An echocardiogram, performed in a local paediatric cardiology service, at the age of 14 months, showed an increase in right ventricular pressure and flow velocity across the conduit. These findings were discussed with our team and due to suspicion of a significant conduit stenosis, the indication for invasive assessment of the conduit and the right ventricular pressure was made.

Cardiac catheterization and bronchoscopy were performed at the age of 18 months. Echocardiography at that time showed an enlarged and hypertrophied right ventricle with normal function. There was moderate conduit stenosis (maximum velocity 3.8 m/s, mean Doppler gradient 25–30 mmHg) and moderate conduit insufficiency.

Bronchoscopy revealed a slit-like tracheal stenosis before the bifurcation and severe left main bronchial stenosis. Cardiac catheterization confirmed the echocardiographic findings of a stenosis of the right ventricular to pulmonary artery conduit with a peak gradient of 30 mmHg and free regurgitation with increased right ventricular pressure. In addition, ectasia of the pulmonary artery bifurcation was demonstrated and thought to be the reason for the bronchial stenosis [Figure 2a and b].

After multidisciplinary team discussion, timely conduit exchange and size reduction of the dilated pulmonary artery to relieve the compressed bronchial structures was decided. For better description of the pulmonary artery anatomy, computed tomography was performed prior to surgery, showing a pulmonary sling with compression of the distal trachea [Figure 3].

Subsequent surgery was performed with change of the right ventricular to pulmonary artery conduit to a 14-mm Contegra graft. Intraoperatively, the sling of the left pulmonary artery was confirmed. The left pulmonary artery took a course from the right side of the chest to the left side of the chest behind the trachea. This was corrected by first explantation of the left pulmonary artery at the exit. Second, the left pulmonary artery was mobilized and extracted from behind the trachea and finally re-anastomosed into the pulmonary trunk. No interventions for the airway stenosis were performed. At her last follow-up appointment, the patient was very well, with stridor occurring only during airway infections.

DISCUSSION

A pulmonary sling is often accompanied by tracheobronchial abnormalities and may be associated with congenital heart disease.[3,6] The diagnosis of common arterial trunk and left pulmonary artery sling, however, is extremely rare, with only a few cases reported so far.[4,5] Anagnostopoulos reported a case of a neonate with heterotaxy, common arterial trunk, right aortic arch, left pulmonary artery sling, and long-segment tracheal stenosis with complete tracheal rings and bilateral left-sided bronchi but no atrial isomerism who underwent common arterial trunk and left pulmonary artery sling repair together with tracheoplasty during neonatal life.[4] In a series of 54 pathologic specimens with common arterial trunk, one case with interrupted aortic arch and a left pulmonary sling is described. In this case, the left pulmonary artery was taking its course between an anomalously arising right upper bronchus and intermediate bronchus directly from the trachea and was then running behind the trachea and in front of the esophagus to the left lung.[5]

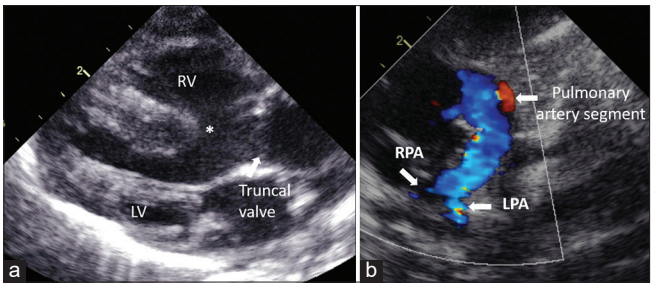

Figure 1: Parasternal echocardiographic views. (a) The arterial trunk is overriding the ventricular septum and the large malaligned ventricular septal (asterisk). (b) After balanced division of the trunk into the systemic and pulmonary pathways, there was a long pulmonary artery segment

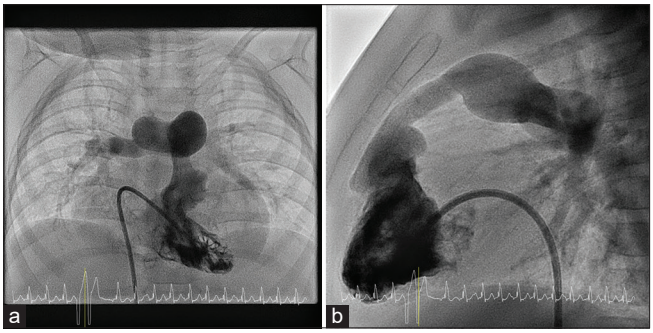

Figure 2: Angiogram in an anterior-posterior (a) and in a lateral projection (b) demonstrating the dilated pulmonary bifurcation and right ventricular to pulmonary artery conduit

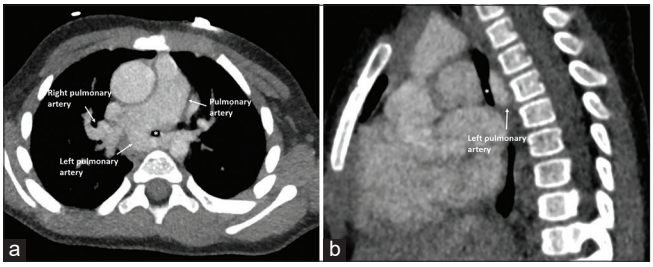

Figure 3: Multislice computed tomography showing the course of the left pulmonary artery (a) and the luminal narrowing of the distal trachea (*, a and b)

Like in our case, the majority of patients with pulmonary sling present early in life, with symptoms ranging from mild stridor to severe airway obstruction.[1] However, in our patient, symptoms were first thought to be related to a dilated pulmonary artery dilatation after common arterial trunk repair and only after a computed tomography scan was performed, a left pulmonary artery sling was diagnosed. Although left pulmonary artery sling in common arterial trunk patients is rare, we therefore recommend that attention should be paid to this condition before and at the time of the initial surgery. Both computed tomography and cardiovascular magnetic resonance are the methods of choice to diagnose pulmonary artery abnormalities, but computed tomography allows better delineation of the tracheobronchial tree.[7]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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