Surgical Repair of Large Pulmonary Artery to Left Atrium Fistula

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Abstract

We report the successful surgical closure of right pulmonary artery to left atrium fistula (RPALAF) with secondary atrial septal defect (ASD) in 3.5 years old girl, who had suffered with central cyanosis and exertional dyspnea.

Key Clinical Message:

We reported a successful surgical closure of right pulmonary artery to left atrium fistula for 3.5 years old girl. The patient's oxygenation and symptoms improved directly after surgery. This case is a typical representation of fistula classification.

Introduction:

The pulmonary fistula is an abnormal connection between the pulmonary artery or its branches and the pulmonary veins or left atrium bypassing the pulmonary capillary bed, which causes a right to left shunt and is considered a rare congenital cardiac malformation (1). Patients suffering from pulmonary fistula have often symptoms of central cyanosis and clubbing, and in case of delayed treatment, patients may develop embolisms, cerebrovascular accidents, or other systemic symptoms (1, 2).

Fewer than 100 cases regarding pulmonary fistulas are published in the medical literature so far (3), and among them there are only 3 published cases of pulmonary artery and left atrium fistulas(4).

This case was difficult to diagnose based on clinical and echocardiographic findings only. Usually, it need a contrast Echo and angiography or a multi-slice CT that is capable of providing the structural anatomy of the right pulmonary artery and its branches and the pulmonary veins and their relationship with the left atrium. It also helps to classify this kind of deformity; hence, we have chosen this appropriate option to repair the deformity. These kind of cases can be manage by surgical closure of the fistula or by device closure using cardiac catheterization (5).

We present here a successful surgical closure of fistula between the right pulmonary artery and the left atrium using a biological patch in a child suffering with central cyanosis.

Case:

A three and half year old girl suffering with central cyanosis since birth, clubbing, exertional fatigue and recurrent respiratory infections was presented. On clinical examination, child was found with severe cyanosis and resting oxygen saturation was 74% in room air. Weight and height of the patient was recorded 13 Kg and 89 cm, respectively. Cardiovascular examination: normal heart sounds with a mild systolic heart murmur on the left sternal border, heart rate 110 beats / minute. The pulse is palpable and symmetrical in the four limbs. Respiratory examination: Clear lungs, pectus excavatum. Examination of the nervous system: normal

The child had frequent respiratory infections and hospitalized due to pneumonia a year ago. Furthermore, laboratory analysis was normal except for HB = 18, ECG: normal. Chest X-ray: Increased vascularity with an opacity most likely representing a vascular silhouette in the right lung.

Echocardiography: Secondary ASD 6.5 mm in diameter with a left to right shunt, enlargement of main pulmonary trunk and the right pulmonary artery, the rest of the echo was normal.Contrast echo confirmed the abnormal connection between the pulmonary arteries and the left atrium through the visualization of bubbles in the left atrium.

Cardiac catheterization was performed, where the presence of the fistula was confirmed between the right pulmonary artery and the left atrium (Fig. 1). Multi-slice CT shows that the pulmonary artery branch supplying the middle right lobe drains directly into the left atrium through a large fistula taking the form of a large aneurysm accompanied by an absence of the upper right pulmonary vein (Fig. 2)

The surgery was performed under general anesthesia through a median sternotomy on a cardiopulmonary bypass, the fistula was found to be 2 - 2.5 cm in size, connecting the right pulmonary artery and the left atrium at the point where the right upper pulmonary vein usually opens into the atrium, the right upper pulmonary vein was absent . By opening the right atrium and through the ASD, the fistula's ostium was reached; closure was performed using a biological patch, followed by closure of the ASD (Fig. 3). Oxygen saturation improved to 95% immediately after surgery, on follow-up. After 6 months, the patient was asymptomatic and the clinical examination was normal.

Discussion:

The clinical findings associated with the occurrence of a direct fistula between the right pulmonary artery and the left atrium. Furthermore, it was vary from mild to extreme cyanosis with clubbing and exertional dyspnea. These patients with above-mentioned symptoms may experience increased blood viscosity, due to the high hemoglobin levels caused by chronic hypoxia. In addition to this, bypassing of the pulmonary vascular bed, the blood filtering function of the lung is lost, which leads to complications such as emboli, brain abscesses, cerebrovascular events and other systemic embolic events (6), one of the rare complications that may occur is fistula rupture.

De Silva et al. (8) described three patterns of fistula between the pulmonary artery and the left atrium depending on where the fistula opened on the left atrium and its relationship with the branches of the right pulmonary artery and pulmonary veins.

Type 1 : The right pulmonary artery and its branches and the pulmonary veins are normal but there is a fistula between the right pulmonary artery and the

left atrium.

Type 2: The lower right pulmonary vein is absent and the lower branch of the right pulmonary artery drains directly to the left atrium where the lower right pulmonary vein usually opens into the atrium.

Type 3: Pulmonary veins drain to an abnormal duct between the left atrium and the right pulmonary artery (a fistula).

Later, the fourth type was added by Oahu et al. 1979 (9). This pattern is similar to the second pattern, where the right pulmonary veins enter the fistula that resembles an aneurysm and connects the lower branch of the right pulmonary vein with the left atrium Fig 4)

In the case we present here, the right pulmonary artery which supplies the middle pulmonary lobe drains directly into the left atrium through a large fistula taking the form of an aneurysm where the upper right pulmonary vein usually drains into the atrium. This case doesn't fall under any particular classification but is somewhat similar to Type-2. (Fig. 5)

There is no definite fetal origin or explanation for the fistula between the right pulmonary artery and the left atrium; some theories suggest the presence of a connection between the right pulmonary artery and the

primary pulmonary vein. At a later stage, the primary pulmonary vein merges into the atrial wall resulting in the fistula (10, 11). Atrial septal defect is the most common cardiac defect associated with the fistula between the right pulmonary artery and the left atrium, the absence of the lower pulmonary lobe and right bronchial atresia are the most common pulmonary malformations (12, 13).

In this case, we found only an ASD of a 6.5 mm diameter that was closed at the surgical repair of the fistula. When diagnosed, this condition should be treated with selective surgery to close the fistula (14). This can be achieved either through outside ligation without cardiopulmonary bypass, or through open heart surgery using a patch which was performed in the case we presented. The surgical approach was selected due the presence of the ASD and the size of the fistula and the thinness of its wall causing concern of rupture. Many other cases were mentioned in the medical literature regarding the closure of the fistula using cardiac catheterization which is an effective alternative to surgical closure.

Conclusion:

In this case, a right pulmonary artery to left atrium fistula was diagnosed as the cause of unexplained cyanosis in 3.5 years old child. The diagnosis was confirmed based on cardiac angiography through cardiac catheterization, and the underlying anatomy of the pulmonary vasculature and its relationship with the left atrium was visualized using multi-slice CT.

This case is of significant importance because classification of fistula does not correspond to the classifications mentioned previously in the medical literature. Surgical repair was performed after diagnosis to prevent the serious complications of the fistula. The patient's oxygenation and symptoms improved immediately after surgery and persisted on follow-up after 6 months.

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Conflict of interest

The authors declare that they have no conflict of interest.

Author contributions

Alaa M Hussain: was responsible for original manuscript writing and editing. Muhamed Younis and Samir Srour: professors who did the surgery and supervising the work. Alaa M Hussain, Mohamed Saleh and Mohammed Ismael: are responsible for data collection, data analysis and manuscript design. Alaa M Hussain: was responsible for manuscript drafting and revision. All authors read and gave the final approval of the manuscript to be published.

Ethical Approval

This case report has been approved ethically by our institution.

Consent for Publication

The authors state that they have written and signed consent from the patient to publish this case report and any accompanying images. A copy of the written consent is available for review by the editor of this journal.

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Figures legends

Fig 1: Diagnosis of the fistula between the right pulmonary artery and the left atrium using angiography catheterization.

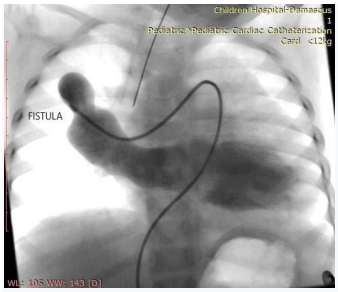
Fig 2: Diagnosis of the fistula between the right pulmonary artery and the left atrium using multislice CT showing the anatomy of the pulmonary vasculature and its relationship with the left atrium through 3D reconstruction.

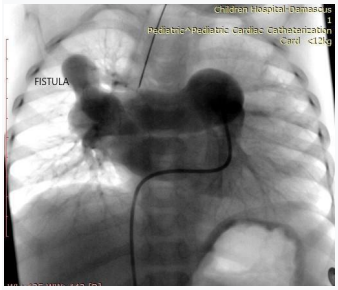
Fig 3: Orifice of the fistula.

Fig 4: Classification of the fistula between the right pulmonary artery and the left atrium.

Fig 5: An illustration demonstrating the fistula between the right pulmonary artery and the left atrium mentioned in this case.

Fig 1





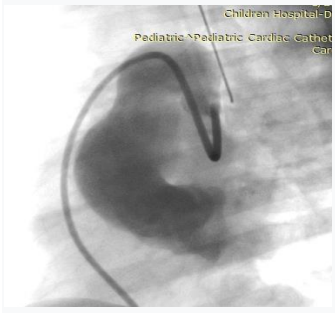


Fig 2