

## Massive left pulmonary artery aneurysm with a co-existing patent ductus arteriosus in a five-year-old female child: A case report

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### Abstract

Pulmonary Artery Aneurysm (PAA), whether congenital or acquired, is a rare diagnostic finding compared to aortic aneurysms. There have been few cases where PAAs were documented as a complication of untreated Patent Ductus Arteriosus (PDA) due to long-standing Pulmonary Arterial Hypertension (PAH). However, it is quite rare for a case of PAA to be reported with co-existing PDA without PAH. This report highlights a case of a five-year-old girl who was presented with palpitations, easy fatigability, fever, cyanosis, and vomiting. A Chest X-ray showed moderate cardiomegaly. A PDA of 6 mm was diagnosed on Transthoracic Echocardiography (TTE) and a large cavity connected with LPA raised suspicion of a possible LPA aneurysm. A Chest CT scan confirmed the diagnosis of a saccular aneurysm, originating from the distal part of the main Left Pulmonary Artery (LPA) just proximal to the point of bifurcation into lobar branches, measuring 7.5x6.5 cm. During surgery, the aneurysm was opened, emptied with suction and closed without resecting the aneurysmal walls. The patient had an uneventful post-op course and is doing well during regular interval follow up visits.

**Keywords:** pulmonary artery aneurysm (PAA), patent ductus arteriosus (PDA), cardiac aneurysm, left pulmonary artery, saccular aneurysm.

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### Introduction

Pulmonary Artery Aneurysm (PAA), either congenital or acquired, is a rare anatomical defect compared to aortic aneurysms; hence, there is a paucity of literature reports.<sup>1</sup> When a true PAA measures more than 5cm (50 mm) in size, it is labelled as a Massive/Giant Pulmonary Artery Aneurysm, which is even more rare.<sup>2</sup>

It is believed that PAA often presents with a spectrum of non-specific symptoms, and the diagnosis is usually incidental.<sup>3</sup> It is most commonly found in the pulmonary trunk and can be classified as peripheral and proximal

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types.<sup>3</sup> Based on the anatomical characteristics of an aneurysm, PAA can also be divided into true (having all three layers of the pulmonary artery involved) and pseudoaneurysms (does not involve true layers of the vessel, instead the wall is made from crosslinks of fibrin/platelets, which is more friable).<sup>3</sup> The aetiology and associations of a PAA usually include congenital cardiac anomalies such as untreated Patent Ductus Arteriosus (PDA) and infective endarteritis.<sup>1</sup>

PAAs are known to occur secondary to a PDA when the treatment is delayed (either by patient's choice or due to delayed diagnosis).<sup>1</sup> This case report highlights the presentation, surgical approach, and outcome of a rare case of PAA with a co-existing PDA.

### Case Report

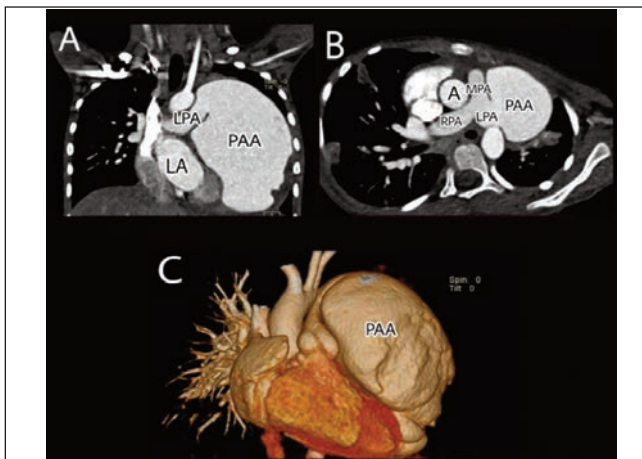
After obtaining consent from the parents of the patient, we are reporting a case of a five-year-old female child, who came to the Paediatric Cardiac Surgery's outpatient department of the National Institute of Cardiovascular Diseases (NICVD), Karachi in May 2022. She presented with symptoms of palpitations, easy fatigability, fever, cyanosis, vomiting, and rashes. These symptoms had been progressively worsening over the past four years. The child's mother described similar complaints dating back to her early infancy with interruption of breastfeeding, diaphoresis, inability to gain weight, and recurrent respiratory tract infections. The child was not undergoing any medical treatment. Her parents did not provide any history of previous hospital admissions, although she had been repeatedly treated for respiratory tract infection on an outpatient basis. Since early infancy, she had been experiencing exertional dyspnoea which worsened over the past three years and she had experienced cyanotic spells for which she had been admitted multiple times, where she was provided supplemental oxygen for a few hours. She responded every time to oxygen therapy, and was sent home but would again develop cyanosis and shortness of breath, repeatedly. The possibility of infective endocarditis was also ruled out based on the patient's history and prior workup.

Upon general physical examination, the patient had a chronically ill appearance and had a Body mass index (BMI)

of 16.8 kg/m<sup>2</sup> (according to the BMI calculator chart of children and teens). Her blood pressures averaged at 110/55 mmHg (Normal blood pressure range in pre-school (3-5 years old) age group=90-120/46-80 mmHg). The character of her peripheral pulses was bounding. On inspection, visible precordial pulsations were noted on the left side of her chest with a point of forceful impulse in the left fifth intercostal space, slightly lateral to the midclavicular line. On auscultation, there was a continuous systolic-diastolic murmur of grade 4/6, audible in the left second intercostal space.

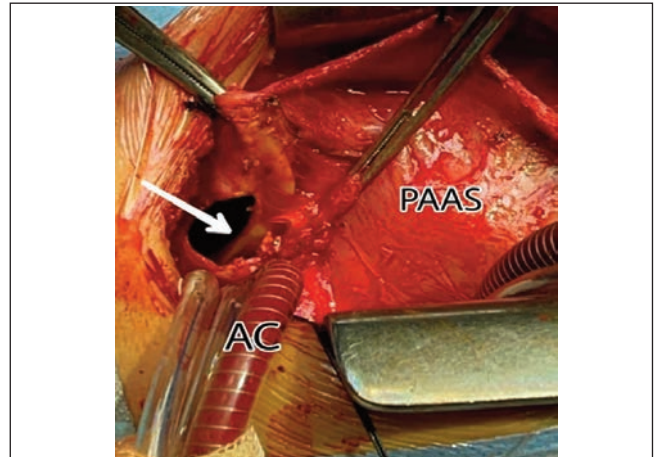
To establish diagnosis, a chest X-ray (CXR), Transthoracic Echocardiogram (TTE), and Computed Tomography (CT) Scan of the chest were advised. The CXR showed moderate cardiomegaly. The TTE revealed a 6 mm PDA with a continuous left-to-right shunt. It also showed that the Left Pulmonary Artery (LPA) was distally connected to a large cavity compressing the left ventricle from behind, raising suspicion of a possible LPA aneurysm. Other findings included EF=60%, TAPSE=17, Main Pulmonary Artery (MPA)=52 mm, Right Pulmonary Artery (RPA)=18.5 mm and LPA=24 mm. There was no significant Pulmonary Arterial Hypertension (PAH) on TTE and all four valves showed no abnormalities. Lastly, a Chest CT scan was done to establish the final diagnosis, which showed a saccular aneurysm originating from the distal part of the main LPA just before the point of bifurcation into lobar branches, which measured 7.5x6.5 cm (75x65 mm), putting it in the category of Massive PAA (Figure 1). The opening of the aneurysmal sac measured 1.4 cm.

Once all the preoperative workup was complete, the



**Figure-1:** Coronal and Axial views of Chest CT Scan (A and B respectively) showing the Massive Pulmonary Artery Aneurysm (PAA). A 3-dimensional CT reconstruction (C) that confirms PAA arising just distal to the left pulmonary artery (LPA).

LPA (Left Pulmonary Artery), RPA (Right Pulmonary Artery), MPA (Main Pulmonary Artery), PAA (Pulmonary Artery Aneurysm), A (Aorta), LA (Left Atrium)



**Figure-2:** Intraoperative photograph, showing pulmonary artery aneurysmal sac (PAAS), aortic cannula (AC), and opening into the aneurysm (arrow).

patient underwent an open PAA repair through a midline sternotomy on the 2022. Moderate adhesiolysis was performed for separating the pericardium. The aneurysm appeared to originate from the distal anterior wall of the LPA, measuring approximately 7.5 cm (75 mm), which hindered proper visualization of the PDA. The patient was taken on cardiopulmonary bypass. Reduced flow was achieved by cardiopulmonary bypass which aided in proper visualization of the PDA. To adequately decompress the pulmonary aneurysm, the duct was properly exposed and double-clamped. While both the ends were quickly repaired following the division of the duct, adequate flows were maintained on bypass. Upon confirming that there were no signs of infection or any other pathology on gross appearance of the aneurysmal sac intra operatively, the aneurysm was opened with a small incision of approximately 2 cm and was emptied via suction (Figure 2). A drain was placed inside the sac without resecting the aneurysmal walls and the opening was stitched. The diagnosis of LPA aneurysm was very typical. Hence, there were no second possibility of any underlying pathology for which a biopsy could have been indicated. The patient was then weaned off bypass and was subsequently extubated in the ICU on the same operative day. The total bypass time was noted to be 15 minutes.

The patient's postoperative stay in the intensive care unit was uneventful. Her CXR was repeated post-op that showed visible anatomical improvement. The patient was called for follow up after 2 weeks of surgery. Later, she used to visit after every 2 to 3 months. Her last follow up was in April 2023 where she seemed to be doing well in terms of her general physical health and investigational parameters.

## Discussion

Pulmonary Artery Aneurysm (PAA) is a rare finding globally, whether congenital or acquired. Most of the diagnosed

cases are of adults who presented later in life with worsening symptoms.<sup>1,2,4</sup> However, there are two cases of a 9-year-old boy and an 8-year-old girl, reported by Rui Huo et al.<sup>4</sup> in 2001, and E Tefera et al.<sup>5</sup> in 2013, respectively, which corresponds to the age group of the patient in our case (five years old) during an early presentation. The diagnosis in all previously reported cases was made by TTE and Chest CT scan, which were the same investigational modalities used in the diagnosis of our patient.<sup>1,2,4,6,7</sup>

When a PAA is diagnosed in a patient (either early or late in life), it is often associated with PDA and PAH.<sup>1</sup> PAH is also reportedly considered a trigger for the development of PAA.<sup>8</sup> One study reports the presence of PAH in 66% cases of PAA.<sup>9</sup> In our patient, TTE confirmed the left-to-right shunt of a PDA with no significant PAH, which correlates with a previous case reported in 2013 by E Tefera and M Teodori.<sup>5</sup> The investigations in our case revealed an aneurysm of the left pulmonary artery, making it a rare and significant finding, as the previous literature states 80% of PAAs to have involved the main pulmonary trunk instead.<sup>10</sup>

Because PAA diagnosis is still in its documenting phase, there are controversies related to its standard treatment. Many interventional procedures, such as a covered stent technique for aneurysm neck exclusion, transcatheter coil embolization, and neck of aneurysm closure via a device have been previously reported to be successful in avoiding surgery for PAA.<sup>11</sup> However, in the case of a PAA rupture, the mortality rate is 100%.<sup>12</sup>

Various factors influence the preference for surgical repair over intervention, such as Massive/Giant PAA, compression of the neighboring great vessels/critical structures, rapidly increasing size, and associated PAH.<sup>4</sup> Surgical approaches vary depending on the case, such as an open surgical repair with or without allogeneic/synthetic grafts.<sup>13</sup> In our patient, open PAA repair was done without graft replacement, as early surgical intervention remains the cornerstone for this disease.<sup>1,14</sup>

## Conclusion

Considering the scarce literature on Pulmonary Artery Aneurysms (PAA), cases like these, reporting Massive PAA with co-existing PDA, are aimed to bridge the knowledge gap worldwide. For the diagnosis of PAAs, a TTE is the first investigation of choice while Chest CT scan remains the gold standard for confirming the diagnosis. TTE can serve as a helpful tool for long-term non-invasive follow-up as well. Co-existing PDA without PAH, highlights the possibility of PAA being congenital. Open surgical repair of Massive/Giant PAA is the cornerstone treatment, as both the outcome and long-term patient prognosis is satisfactory.

**Consent:** The parents of the patient signed a written consent for publishing this case for scientific purposes only, ensuring the patient's anonymity.

**Disclaimer:** The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

**Conflicts of Interest:** None.

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**Author Contribution:**

JK: Conceived the idea, designed the manuscript, responsible for the integrity of research.

MMK: Manuscript writing, drafting and editing.

ZB, MM, SKB: Review and final approval of the manuscript.