Annals of International Medical and Dental Research
E-ISSN: 2395-2822 | P-ISSN: 2395-2814
Vol-10, Issue-1 | Jan-Feb 2024
https://doi.org/10.53339/aimdr.2023.10.1.12
Page no- 98-105 | Section- Research Article (Cardiac Surgery)

A Rare Tetralogy of Fallot patient with Absent Left Pulmonary Artery Syndrome Successfully Treated in BSMMU Cardiac Surgery- A Case Report

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Received: 01 October 2023
Revised: 30 October 2023
Accepted: 13 November 2023
Published: 31 December 2023

Abstract

Congenital unilateral absence of a pulmonary artery (UAPA) is a rare anomaly. The absence of right pulmonary artery is usually not associated with cardiac diseases but the absence of left pulmonary artery (LPA) is associated with cardiac diseases. Absent LPA is usually associated with tetralogy of Fallot (TOF) and hence surgically treated. As such, the agenesis of the unilateral pulmonary artery without any cardiovascular malformation is rare. This malformation of the absence of left pulmonary artery is confirmed by cardiac catheterization and CT angiography, prior to this X-ray gives some clues regarding the diminished vascularity of the involved left lung. The incidence is approximately 1 in 2,00,000 individuals with diverse nonspecific clinical presentation. Chest X-ray and echocardiography are usually the initial supportive investigations. Chest X-ray may reveal a minimally small left pulmonary hilum and left lung. MDCT angiography of pulmonary arteries may reveal an absent left main pulmonary artery with systemic collaterals around the left hemithorax. An early diagnosis of a unilateral absence of pulmonary artery and appropriate intervention has a beneficial role in improving the outcome. Diagnosis can be hard due to the rarity of the circumstance and its nonspecific presentation. We are hereby reporting a case of absent left pulmonary artery associated with tetralogy of Fallot which was incidental finding during evaluation of cyanotic patient. We did successful intracardiac repair with good outcome and without any major adverse effect.

Keywords:- Pulmonary Artery Syndrome, Unilateral Absence Of A Pulmonary Artery, Tetralogy Of Fallot

INTRODUCTION

The congenital unilateral absence of the pulmonary artery (UAPA) is a rare anomaly that is frequently associated with other cardiovascular abnormalities.[1] Agenesis of the unilateral absence of the pulmonary artery (UAPA) generally occurs in patients with situs solitus and concordant atrioventricular and ventriculoarterial connections.[2] According to Kucera and colleagues,[3] the incidence of UAPA is 0.6%. Forty percent of patients have

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the isolated form of lesion, whereas in 60% of cases, UAPA is combined with other congenital heart defects. Tetralogy of Fallot is the most frequent concomitant disorder for UAPA.[3] From 1% to 3% of patients with tetralogy of Fallot have UAPA. Interestingly enough, the left pulmonary artery is absent five to eight times more frequently than the right one.[4] According to the theory of involution of the sixth aortic arch, the absorption of either the right or left arch leads to the absence of one of the main pulmonary branches.[5] The concept, however, does not explain the frequent association of agenesis of the left pulmonary artery with tetralogy of Fallot or why the absence of the right pulmonary artery is usually not associated with other congenital heart defects. According to the ontogenetic theory, the absence of one of the pulmonary arteries is caused by abnormal septation of the truncus Dorsal shift of either the right or left truncal ridges results in agenesis of the right or left pulmonary arteries. The theory successfully explains the fact that agenesis of the right pulmonary artery is most often an isolated anomaly as well as why a frequent association of tetralogy of Fallot with the absence of the left pulmonary artery does occur.[6] World literature describes fewer than 100 cases of surgical treatment of tetralogy of Fallot associated with UAPA. Palliative surgery encounters approximately 20 operations with a high hospital mortality rate of 35%.[7] The number of complete repairs of tetralogy of Fallot with UAPA does not exceed 70 operations with hospital mortality of approximately 8% (Liu et al., 1984).[8] The first reports of successful complete repair of tetralogy of Fallot with concomitant absence of the left or right pulmonary artery were published in 1962 and 1975, respectively.[9] However, this type of intervention still represents a surgical challenge and is associated with a relatively high operative risk. A certain percentage of patients require heart or heart lung transplantation after a complete repair because of biventricular failure. The aim of the present study is to summarize our 22-year experience in surgical correction of tetralogy of Fallot associated with UAPA as well as to present novel approaches to the treatment of this complex disorder.[10]

We are reporting a rare case of an 8-year-old boy admitted to Cardiac surgery at Bangabandhu Sheikh Mujib Medical University who presented with shortness of breath on exertion and chronic cough since childhood and was diagnosed as a case of left pulmonary artery agenesis with other cardiac anomalies, tetralogy of Fallot.

CASE REPORT

An eight (8) years old male child, the first issue of non-consanguineous parents, immunized as per EPI schedule was admitted in cardiac surgery of BSMMU with complaints of continued fever, cough, and difficulty in breathing for 2 days. He also had bluish discoloration of his tongue and finger since his birth and failed to thrive. He also gives a history of recurrent respiratory tract infections for which he was being conservatively managed previously. Later he was referred to our cardiac surgery department for definitive management of his condition.

On examination, he was looking unwell, alert, conscious, and cooperative patient. He was found cyanotic both central and peripheral and clubbing was present on both upper and lower

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limb fingers and toes. There was normal jugular venous pressure, no edema, anaemia and jaundice was absent. Patient height was 110cm and weight-16kg, Pulse-92/min, regular, BP-90/60mmHg, R/R-32/min. On precordium examination, apex beat was thursting in nature and systolic thrill was present more prominent in pulmonary area. Harsh ejection pan systolic murmur was present in the pulmonary area. ECG showed sinus rhythm and right ventricular hypertrophy (RVH) (Figure-1). His chest x-ray (Figure-5) showed boot shaped heart. His echocardiogram revealed large malaligned VSD with 40-50% overriding of aorta with bidirectional shunt mostly right to left shunt, severe infundibular and valvular pulmonary stenosis (Figure-2). Normal Pulmonary Artery pressure and good biventricular function, right atrium (RA) and right ventricle (RV) dilated, RV is hypertrophied and trabeculated. Color Doppler showed PV: Velocity-4.5 m/sec, PPG-84mmHg. LA-19 mm, AO 25mm. His CTangiogram (Figure 3 and 4) showed no persistent superior venacava (PLSVC) and RV graphy showed thickly trabeculated, hypertrophied and normally functioning RV ejecting to PA. Critical infundibular and valvular PS noticed. Good main pulmonary artery (MPA) and right pulmonary artery (RPA) anatomy. Left pulmonary artery (LPA) is not seen in the anteroposterior shadow left anterior oblique (LAO) view. RPA: 12.5mm, LPA-absent. Descending aorta at the level of the diaphragm: 12.65mm. Normal pulmonary venous drainage and small mediastinal aortopulmonary collaterals. LV angiography showed smooth wall LV ejecting to Aorta. A large perimembranous VSD was noticed. Normal origin of epicardial coronary arteries. The aortogram showed a left aortic arch, no PDA, and no MAPCA. Cardiac catheterization done and the diagnosis was tetralogy of Fallot (TOF) with absent LPA syndrome. Other Investigations showed Hb-18.5 gm/dl, Total White Cell count- 6,480/mm3, Neutrophil- 47%, Lymphocyte-43%, Platelet-3,40,000, ESR-13mm/1St hr, BT-3min30s, CT-5min30s, ALT-26 U/L, S. Creatinine - 0.62mg/dl, PT-11.7, INR-0.97, CRP-1.8 mg/l. After confirming the diagnosis we planned for surgery.

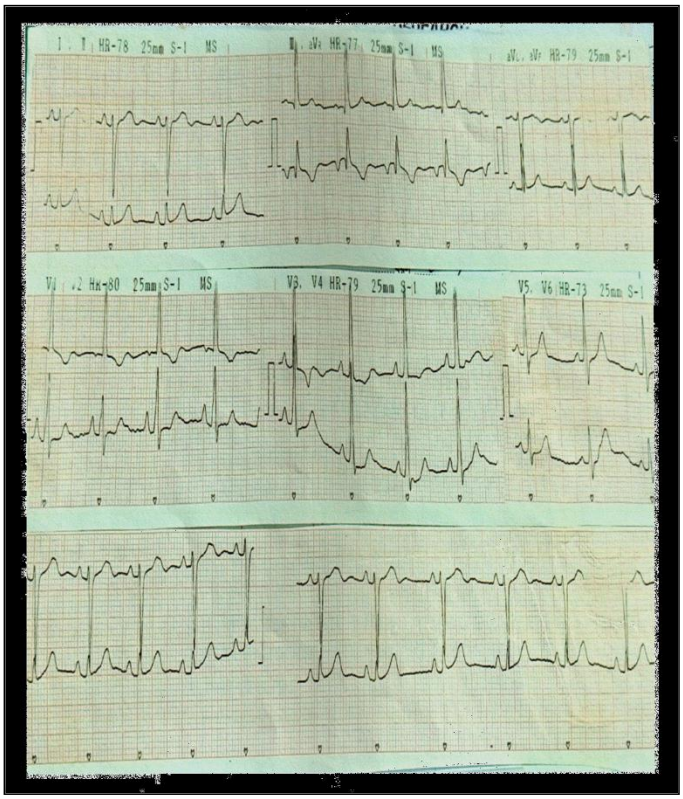

Figure 1: ECG showed sinus rhythm and right ventricular hypertrophy (RVH).

Under all aseptic precautions, a standard midline sternotomy was done. Cardiopulmonary bypass was established. Cardiac arrest was achieved by antegrade St. Thomas cardioplegia. Right atriotomy done and

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anatomy assessed. The infundibular muscle band was resected. Main Pulmonary Artery (MPA) augmentation done with a pericardial patch. VSD was identified and repaired with a polytetrafluroethylene (PTFE) patch. RA tomy was closed in layers with one RV epicardial pacing wire and two chest drains kept in right pleural and mediastinal cavity respectively. After proper hemostasis, the wound was closed in layers. Aortic cross clamp time was 96 minutes and cardiopulmonary bypass time-165 minutes. Extubation was done in the usual time duration. His postoperative period was uneventful. He was discharged with medication on this 9th postoperative day with the advice of further follow-up. Patient came for followup after 15 days and his wound was found healthy and healing well, no dischage or collection in addition sternum was stable.

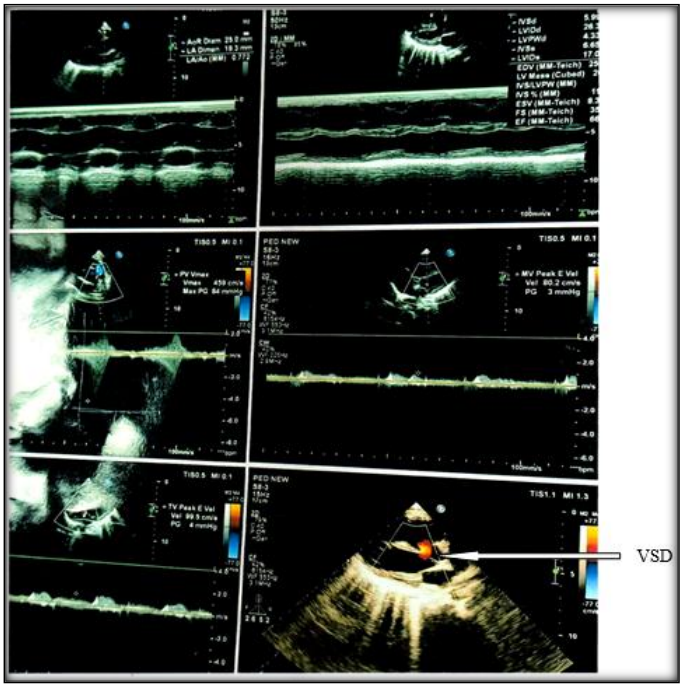

Figure 2: Echocardiogram showing large malaligned VSD with 40-50% overriding of aorta with bidirectional shunt mostly right to left shunt, severe infundibular and valvular pulmonary stenosis.

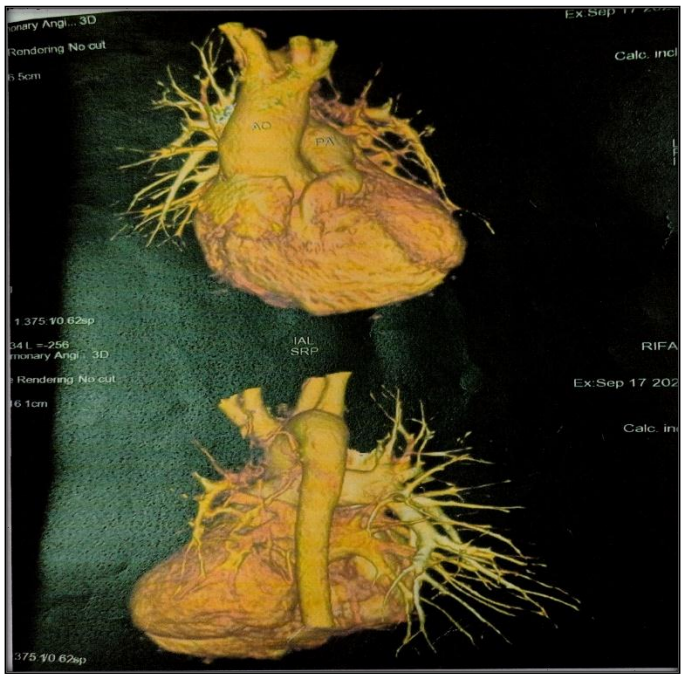

Figure 3: CTangiogram of heart and great vessels showing absent left pulmonary artery.

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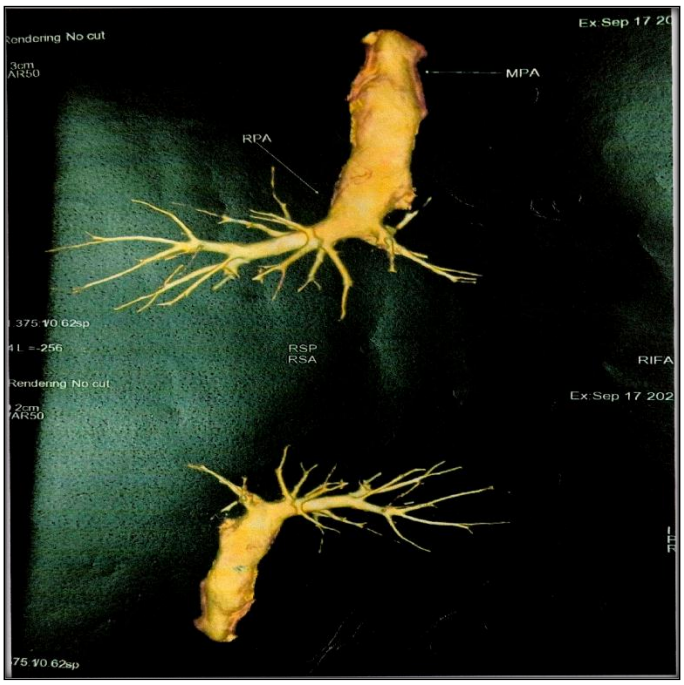

Figure 4: CTangiogram of heart and great vessels showing absent left pulmonary artery.

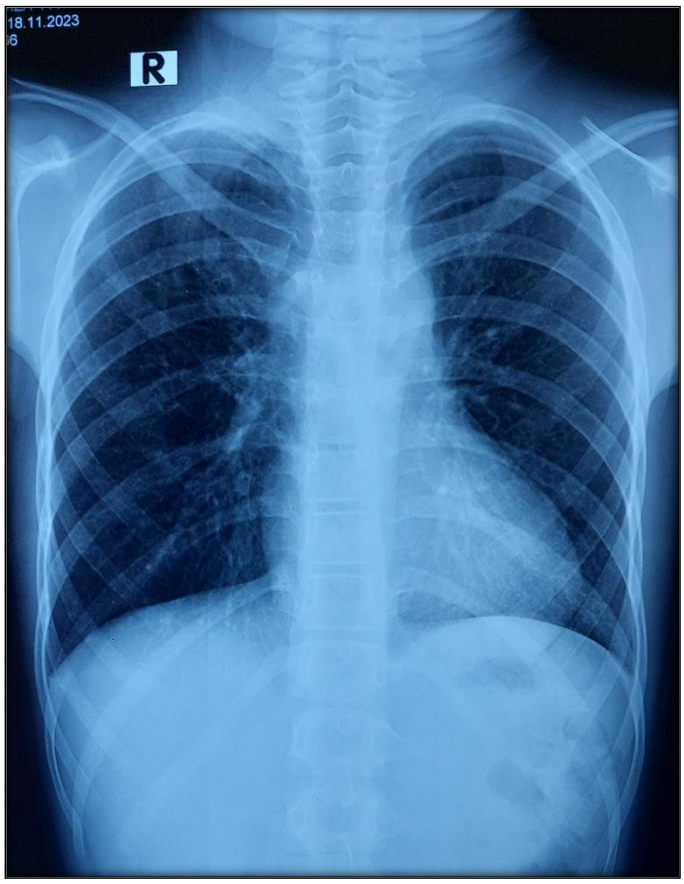

Figure 5: Chest X-ray P/A view showing boot shaped heart.

DISCUSSION

UAPA is a rare congenital vascular anomaly usually coexists along with other cardiovascular abnormalities. It has been observed that distal intrapulmonary branches of the affected artery are usually intact and may be supplied by collateral vessels from bronchial, intercostal, internal mammary, subdiaphragmatic, subclavian, or even coronary arteries.[11] UAPA may also occur as an isolated finding. Unilateral absence of pulmonary artery is usually treated surgically during the first year of life. Isolated UAPA generally has a mild clinical course, few may have pulmonary hypertension.[12] Highly

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developed collaterals leading to recurrent hemoptysis demand embolization of the bronchial arteries, so perioperative bleeding from the collateral vessels like bronchial, intercostal, subclavian or sub diaphragmatic arteries is prevented.[13]

In 1868, Fraentzel described the first case of Isolated UAPA.[14] The most common symptoms of UAPA are recurrent pulmonary infections, dyspnea, and, less frequently, hemoptysis and signs of pulmonary hypertension,12 while 13%—15% of UAPA are asymptomatic.[1] In our patient also have the recurrent respiratory tract infection, dyspnea, cough which is consistent with the previous study by Harkel et al.[12] However, isolated UAPA patients may complains of recurrent hemoptysis or recurrent pulmonary infections require surgical intervention.[13] Pulmonary hypertension is due to an increased blood flow in the contralateral pulmonary artery leading to remodeling of the arterioles which later offers increased vascular resistance.[12]

The diagnosis of isolated UAPA is a big challenge because of subtle signs.[1] An ejection systolic murmur across the pulmonary outflow tract could be found. The electrocardiogram is usually normal while transthoracic color doppler echocardiography is a useful tool for UAPA and the associated anomalies. Imaging studies, MRI, or computed tomography arteriography is required for confirmation of the diagnosis and delineates pulmonary vasculature. Chest radiography and MRI may show ipsilateral small hemithorax, mediastinal with tracheal shift toward the ipsilateral side, ipsilateral hemidiaphragm elevation, ipsilateral diminished pulmonary vascular markings, and contralateral lung hyperinflation.[1] Although our patient had only boot shaped heart in chest x-ray but there was no tracheal shifting or small hemithorax.

However, in regard to the adequate treatment for isolated UAPA, there is no consensus yet, and has been said that it depends on symptoms, collateral vasculatures and the anatomy of pulmonary artery. Regular follow-up to monitor the development of pulmonary hypertension is also needed. In the case of pulmonary hypertension, pulmonary artery dilating treatment viz endothelin receptor antagonist (i.e. Bosentan), prostacyclin, and nitric oxide may be beneficial. Surgical correction treatment to augment anterograde blood flow to the affected lung or lobectomy is practiced.[1]

The treatment objective of UAPA is to correct intracardiac defects and also ensure physiologic blood flow in the ipsilateral lung especially performed for isolated UAPA.[1] The rationale of the complex staged procedure is that lobar, segmental, and acinar pulmonary arteries of the ipsilateral lung may help preservation of normal structure and function. Pulmonary artery continuity is established with a conduit however adequate size of the pulmonary hilar artery is the prerequisite for successful repair and establishing proper pulmonary circulation.16 In our case we did straight forward excision of infundibular muscle band and vulvotomy with transannular augmentation by pericardial patch and PTFE patch closure of VSD. We did not do anything for the left lung. Patient outcome was good with no major adverse cardiovascular event which showed consistent result with the study done by Bockeria et al 2013.[17] A systemic-to-pulmonary shunt as a palliative procedure is done for

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tetralogy of Fallot associated with UAPA.16 Systemic to pulmonary shunt in UAPA is associated with high operative risk and hence transventricular infundibulectomy and valvotomy may also be better options.[17,18]

CONCLUSIONS

Transthoracic color Doppler echocardiography is the first-line diagnostic tool, and chest CT angiogram and MRI help in the correct diagnosis of UAPA associated with cardiac anomalies. It further helps to identify collateral arteries of the ipsilateral lung. Early diagnosis and regular follow-up are necessary for better medical therapy or surgical repair of this defect. However, late diagnosis of UAPA delays the age at presentation, and henceforth higher risk of morbidity and mortality and greater treatment hindrances.

REFERENCES

1. Kruzliak P, Syamasundar RP, Novak M, Pechanova O, Kovacova G. Unilateral absence of pulmonary artery: pathophysiology, symptoms, diagnosis and current treatment. Arch Cardiovasc Dis. 2013;106(8-9):448-54. doi: 10.1016/j.acvd.2013.05.004.
2. Bockeria LA, Podzolkov VP, Makhachev OA, Zelenikin MA, Alekian BG, Ilyin VN, et al. Surgical correction of tetralogy of Fallot with unilateral absence of pulmonary artery. Ann Thorac Surg. 2007;83(2):613-8. doi: 10.1016/j.athoracsur.2006.08.022.
3. Kucera V, Fiser B, Tůma S, Hucin B. Unilateral absence of pulmonary artery: a report on 19 selected clinical cases. Thorac Cardiovasc Surg. 1982;30(3):152-8.
4. Zhang GC, Wang ZW, Zhang RF, Zhu HY, Yi DH. Surgical repair of patients with tetralogy of Fallot and unilateral absence of pulmonary artery. Ann Thorac Surg. 1997;64(4):1150-3. doi: 10.1016/s0003-4975(97)00822-9.
5. Sherrick DW, Kincaid OW, Dushane JW. Agenesis of a main branch of the pulmonary artery. Am J Roentgenol Radium Ther Nucl Med. 1962;87:917-28.
6. Cucci CE, Doyle EF, Lewis EW Jr. Absence of a primary division of the pulmonary trunk. An ontogenetic theory: An ontogenetic theory. Circulation. 1964;29(1):124–31.
7. Gamba A, Villani M, Tiraboschi R, Annecchino P, Crupi G, Vanini V, et al. Surgical treatment of the tetralogy of Fallot with a single pulmonary artery. G Ital Cardiol. 1984;14(7):499–504.
8. Liu JS, Wang ZW. Correction of tetralogy of Fallot in patients with a single pulmonary artery. Zhonghua Wai Ke Za Zhi. 1994;32(3):172–4.
9. Turinetto B, Colì G, Donati A, Galli R, Mikus P, Pierangeli A. Absent right pulmonary artery complicating Tetralogy of Fallot. J Cardiovasc Surg (Torino). 1975;16(3):322–6.
10. Lamour JM, Hsu DT, Quaegebeur JM, Pinney SP, Mital SR, Mosca RS, et al. Heart transplantation to a physiologic single lung in patients with congenital heart disease. J Heart Lung Transplant. 2004;23(8):948-53. doi: 10.1016/j.healun.2004.06.012.
11. Shamsuddin AK, Biswas PK, Manzoo MISA, Azad MAK, Hasan MNA, Hosain J, et al. Tetralogy of Fallot with Absent Pulmonary Valve Syndrome with Absent Left Pulmonary Artery - A Rare Presentation. Bangladesh Heart J. 2020;35(1):74-7.
12. Ten Harkel AD, Blom NA, Ottenkamp J. Isolated unilateral absence of a pulmonary artery: a case report and review of the literature. Chest. 2002;122(4):1471-7. doi: 10.1378/chest.122.4.1471.
13. Takahashi T, Endo H, Ito T, Takei T, Yagi K. Isolated unilateral absence of the left pulmonary artery: a case report. Ann Vasc Dis. 2014;7(2):178-82. doi: 10.3400/avd.cr.14-00005.
14. Cui M, Xia B, Wang H, Liu H, Yin X. A Rare Case of Adult Aortopulmonary Window Combined with Anomalous Origin of the Right Pulmonary Artery from the Aorta Leading to Eisenmenger Syndrome. J Int Med Res. 2021;49(1):300060520984656. doi: 10.1177/0300060520984656.
15. Kim KW, Lee JI, Son KH, Kim EY, Park KY, Park CH. Isolated Unilateral Absence of Pulmonary Artery

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Page no- 98-105 | Section- Research Article (Cardiac Surgery)

Associated with Contralateral Lung Cancer. Korean J Thorac Cardiovasc Surg. 2018;51(4):280-282. doi: 10.5090/kjtcs.2018.51.4.280.
16. Kosaka Y, Kurosawa H, Hoshino S, Shin'oka T, Isomatsu Y, Tsuji Y. Surgery for unilateral absence of pulmonary artery using autologous tissue. Ann Thorac Surg. 2003;76(4):1281-3. doi: 10.1016/s0003-4975(03)00521-6.
17. Bockeria LA, Podzolkov VP, Makhachev OA, Alekyan BG, Khiriev TKh, Zelenikin MA, et al. Palliative surgical treatment of congenital heart defects associated with unilateral absence of the pulmonary artery. Interact Cardiovasc Thorac Surg. 2013;16(3):286-92. doi: 10.1093/icvts/ivs503.
18. Goldsmith M, Farina MA, Shaher RM. Tetralogy of Fallot with atresia of the left pulmonary artery: surgical repair using a homograft aortic valve. J Thorac Cardiovasc Surg. 1975;69(3):458-66.

Source of Support: Nil, Conflict of Interest: None declared