Acta Biomed 2025; Vol. 96, N. 2:16652 DOI: 10.23750/abm.v96i2.16652

Case report

Successful pulmonary arteriovenous malformation closure using MFO in child

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Abstract. *Background and aim:* Pulmonary arteriovenous malformations (PAVM) in children is a cardiovascular anomaly characterized by an anatomic communication between the pulmonary artery and pulmonary vein. PAVM is a rare disease, reported more in females. All PAVMs of size at least 3 mm have to be treated to prevent lethal complications such as stroke/brain abscess. *Methods:* A 3 years old boy weighing 13 kgs experienced easy fatiguability, exertional dyspnea NYHA class II, and delayed milestone. Peripheral oxygen saturation was 72%. Under general anesthesia, a 6French introducer sheath was placed in the right common femoral vein. The right heart was catheterized with a 5F pigtail directed engaged the right pulmonary artery, and pulmonary angiogram showed a connection between pulmonary artery and pulmonary vein. The target vessels for occlusion were the superior (7.6 mm) and inferior (7.7 mm). A 12-10 Konar MFO device was positioned to occlude the connection. *Results:* Pulmonary angiography showed no residual flow, peripheral oxygen saturation improved significantly to 92% after occlusion. *Conclusions:* This case demonstrates the feasibility of percutaneous closure of giant PAVM with good result. No mortality or complication were reported up to 4.5 years post procedure. (www.actabiomedica.it)

Key words: pulmonary arteriovenous malformation, transcatheter closure, konar-mf device, hypoxemia

Introduction

Pulmonary arteriovenous malformations (PAVMs) are described as rare vascular anomalies characterized by abnormal connections between pulmonary arteries and veins, bypassing the normal capillary bed and leading to right-to-left shunting. The occurrence of PAVMs is estimated to be between 2 to 3 cases per 100,000 individuals in the general population, with a notable prevalence in pediatric populations, where approximately 10% of cases are diagnosed in children (1,2). In children, PAVMs can manifest as solitary or multiple lesions, with solitary PAVMs occurring in 42% to 72% of cases (1,2). Furthermore, it can cause severe morbidity & mortality by occurrence of complications like cerebral vascular accident, brain abscess, haemothorax, life-threatening haemoptysis if not treated. The prevalence of pulmonary arteriovenous malformations (PAVMs) in children is expected to be around 1 in 2,600, a figure that reflects the significant increase in detection rates due to advances in diagnostic technologies including CT angiography (3). Although most children with pulmonary arteriovenous malformations (PAVMs) remain asymptomatic (4), this condition can lead to serious complications, including hypoxemia, stroke, and massive hemoptysis (5). Other common complications include paradoxical embolism, which may result in stroke and cerebral abscess (5), respiratory disorders such as hypoxemia and respiratory failure (6), as well as cardiovascular risks including myocardial infarction and arrhythmias (5). Initial screening and management is of utmost importance, especially in children with hereditary hemorrhagic telangiectasia (HHT), in light of the potential for severe

complications (7). Yet, the dearth of this condition can often lead to underdiagnosis, increasing the urgency for awareness among medical personnel (3,8).

Case report

A 3-year-old boy presented to pediatric emergency room with complaints of shortness of breath. The parents said that their child get tired easily, especially in the last 6 months, had dyspnea during activities, and unable to walk for a long time. Gross motor, speech and personal social development were delayed. He reported no issues with coughing, fever, body swelling, chest pain, or seizures and squatting. There was no history of congenital heart disease, lung disease (such as tuberculosis), or cancer in the family. In this study, we utilized medical consent documents, a concept implemented by granting guardians or parents the right to consent to medical procedures for patients who lack the legal capacity to provide consent themselves, as indicated in the section governing patient/guardian signatures. In principle, parent-informed consent refers to the consent given by parents or legal guardians after receiving adequate information about the benefits, risks, and alternatives of the medical procedure to be performed. In the documents we used, the key elements of informed consent such as information about risks, benefits, and the right to ask questions were clearly stated and aligned with applicable ethical standards and regulations. During the physical examination, the patient appeared malnourished with signs of central cyanosis, conjunctival congestion, and retraction of the chest and nasal breath. Auscultation showed bilateral vesicular breath sounds, absent of crepitation or rhonchi; heart sounds were normal as well, with no discernible murmur. The examination of the abdominal and nervous systems was normal. The peripheral oxygen saturation ranged from 69% to 75%, accompanied by finger clubbing. Laboratory tests reveal hemoglobin at 17 gm/dL, hematocrit at 54%, and platelets at 344,000. Kidney and liver function tests were normal. Chest X-ray indicated opacity in the right lower zone of the lung (Figure 1), while transthoracic echocardiography appeared normal. Based on the parents' accounts and corroborated by physical

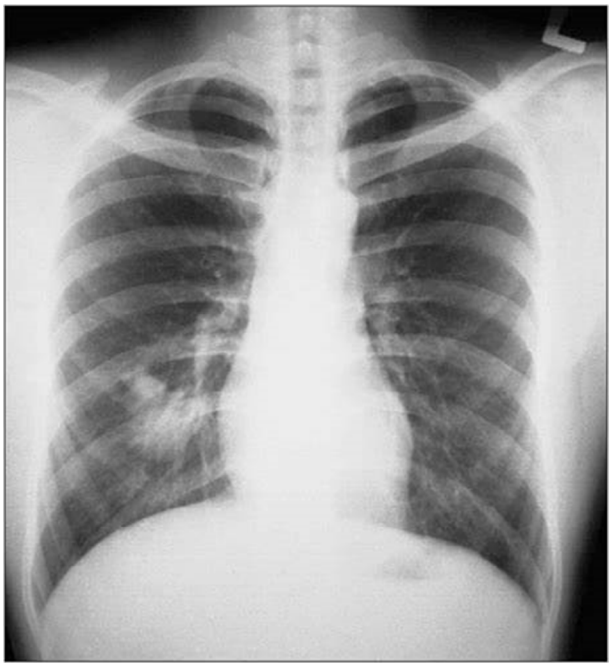

Figure 1. Chest X-Ray of the patient.

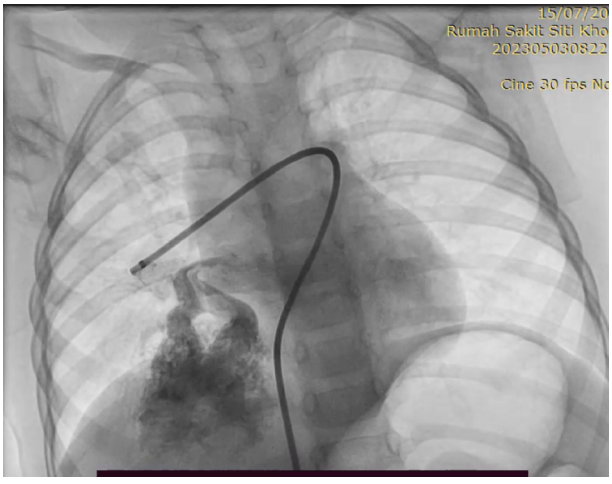

Figure 2. Connection between pulmonary artery and vein, with size 7,7 mm.

and lab results, cyanotic heart disease was suspected, leading to the patient's scheduling for heart catheterization. Right pulmonary artery angiography was performed, revealing contrast filling the right pulmonary artery, passing to the right pulmonary vein and entering the left atrium, with a size of approximately 7.6-7.7 mm (Figure 2). Left pulmonary artery angiography

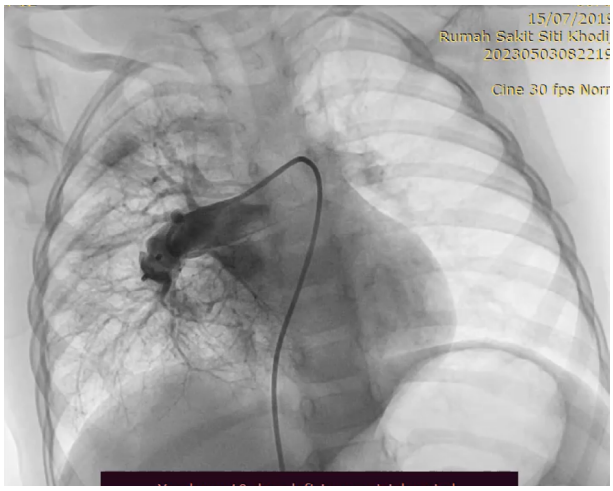

Figure 3. Post occlude closure showed no residual.

was normal. The procedure for closing the pAVM was carried out using a Lifetech Konar-MF device sized 12-10. Under fluoroscopic guidance, a femoral venous approach was used to gain access. A catheter was advanced to the pulmonary artery, and selective angiography was performed to identify the pAVM location and dimensions. Following this, the Lifetech Konar-MF device was deployed through the catheter system, ensuring precise positioning across the AVM. The deployment process required careful manipulation to avoid dislodgement or incomplete occlusion, as the shunt location posed a challenge in ensuring device stability. Angiography post-embolization confirmed no residual shunt and successful occlusion of the pAVM by the device (Figure 3). No procedural complications, such as device migration or vascular injury, were observed. Follow-up evaluations at 3 and 6 months demonstrated favorable outcomes. Vital signs were stable, with no evidence of hypoxemia or recurrent symptoms. Echocardiographic assessments were performed during each follow-up visit to evaluate cardiac function and confirm the absence of residual shunting or device-related complications. The echocardiograms showed no abnormalities, further corroborating the successful closure of the pAVM. These findings underscore the efficacy of the intervention in achieving complete occlusion and favorable clinical outcomes.

As described in Table 1, the chronology of the patient's diagnosis and treatment started from the initial complaint, diagnostic examination, to long-term follow-up. At the initial stage, the patient showed symptoms such as shortness of breath, fatigue, central cyanosis, and low oxygen saturation (69-75%). Subsequently, various diagnostic tests, including chest X-ray and ECHO, are performed to identify potential cardiovascular problems. Based on the results of these tests, the medical team suspected cyanotic heart disease.

The table also notes notable interventions such as percutaneous embolization that successfully removed a pulmonary arteriovenous shunt (pAVM), followed by an increase in oxygen saturation to 98% after the procedure. Post-action monitoring showed continued improvement, with 3-month and 6-month follow-up examination results showing normal vital signs and improved physical activity tolerance.

Discussion

Pulmonary arteriovenous malformations (PAVMs) are irregular vascular formations marked by direct links between pulmonary arteries and veins, circumventing the standard pulmonary capillary network. This unique pathophysiology results in a right-to-left shunt, which can lead to significant clinical consequences, including hypoxemia and paradoxical embolism. The development of PAVMs can be congenital or acquired, with congenital cases being more prevalent in pediatric populations (9). The pathophysiology of PAVMs involves a failure in the normal development of the pulmonary vasculature, which can be attributed to genetic factors, particularly in conditions such as hereditary hemorrhagic telangiectasia (HHT). In HHT, mutations in genes responsible for vascular development, which could lead to the formation of abnormal blood vessels, increasing the risk of PAVMs. Approximately 5-15% of patients with HHT develop PAVMs, which can manifest as a significant cause of morbidity due to the associated risks of hypoxemia and embolic events (10,11). In children, PAVMs can also arise as a complication of certain surgical procedures, such as the Kawashima operation or the Fontan procedure, which alter the hemodynamics of the pulmonary circulation. These surgeries could lead to the development of PAVMs due to the absence of normal pulmonary vasculature

Table 1. Timeline of Patient Diagnosis and Treatment.

Timeframe	Event Description	Diagnostic Tests Performed	Findings/Results	Interventions/Outcomes
Presentation	Shortness of breath, fatigue, delayed development, central cyanosis, low oxygen saturation (69–75%).	Clinical examination, laboratory tests, chest X-ray	Central cyanosis, finger clubbing, opacity in right lung zone.	Scheduled for cardiac catheterization.
Day 1	Initial diagnostic tests performed.	Chest X-ray, ECHO	Chest X-ray: opacity in right lung zone. ECHO: normal.	Diagnosis of suspected cyanotic heart disease.
Day 2	Right pulmonary artery angiography performed.	Pulmonary angiography	Identified pAVM (7.6-7.7 mm) in the right pulmonary artery.	Decision to proceed with percutaneous embolization.
Day 3	Percutaneous embolization of pAVM.	Post-embolization angiography	Complete occlusion of pAVM, no residual shunt.	Peripheral oxygen saturation increased to 98% with oxygen support.
Post-procedure (24 hours)	Recovery phase and monitoring.	Oxygen saturation monitoring	Cyanosis and dyspnea resolved; oxygen saturation 96% (room air).	Patient discharged with follow-up plan.
Follow-up (3 months)	Routine check-up and evaluation.	ECHO, clinical examination	Normal vital signs, no residual shunt.	Continued improvement in physical activity tolerance.
Follow-up (6 months)	Routine check-up and evaluation.	ECHO, clinical examination	Normal vital signs, no residual shunt.	Patient demonstrated sustained clinical improvement.

development. Due to that, transcatheter occlusion has emerged as a preferred treatment modality considering its minimally invasive nature and effectiveness. The occlusion of these malformations is crucial in preventing such complications, particularly in pediatric patients who may present with severe symptoms like cyanosis and exercise intolerance (12,13). Embolization therapy is a treatment method that involves blocking the feeding arteries, including the use of Amplatzer vascular plugs. Embolization therapy removes the necessity for surgical procedures and results in lower morbidity and mortality (14). Transcatheter occlusion involves the use of various devices, such as coils and vascular plugs, to close the abnormal connections. The Amplatzer Vascular Plug (AVP) has been noted for its effectiveness in embolizing PAVMs, allowing for a targeted approach to occlusion without the need for surgical intervention (15). This technique is particularly advantageous in children, as it minimizes recovery time and associated risks compared to traditional surgical methods. Studies have shown that endovascular embolization can achieve high rates of complete occlusion, with low rates of complications (16). In this case we use Konar-MF to occlude PAVM. This device in similar to vascular plug, with two terminal disks, the use of these device for fistula embolization expands the options of available material. The patient showed significant improvement in dyspnea, cyanosis, and fatigue after undergoing embolization therapy, avoiding complications from PAVM and enhancing their quality of life without morbidity.

This study emphasizes the importance of transcatheter interventions for closing pulmonary arteriovenous malformations (PAVM) in pediatric patients, as previously reported in various case studies. For example, Beck et al. documented the effectiveness of using Amplatzer devices in closing PAVM in patients of different ages with a high success rate and minimal serious complications (14). Furthermore, the study conducted by Shimohira et al. highlights the role of embolization utilizing modern devices in the management of PAVM, which has shown promising

clinical outcomes with minimal risks (16). The cases we reported provide clinical evidence that the Konar-MF device could be a highly effective alternative, with clinical outcomes equal to or even better than more commonly used devices like the Amplatzer Vascular Plug. This finding is in line with literature indicating that minimally invasive interventions can lead to significant improvements in patients with complex vascular complications. This case highlights a distinctive contribution to the existing literature on pulmonary arteriovenous malformations (PAVMs) through the application of the Konar-MF device for effective occlusion in a pediatric patient, a scenario that has not been extensively documented. While embolization therapy, particularly utilizing devices like the Amplatzer Vascular Plug, is well-established, the Konar-MF device presents a novel alternative with its unique dual terminal disc design, representing an innovative advancement in the field of PAVM occlusion. Furthermore, the case illustrated significant clinical improvements, including the resolution of cyanosis and dyspnea following the procedure, aligning with the anticipated benefits of transcatheter embolization. The patient's swift recovery and the minimal morbidity experienced post-procedure underscore the increasing efficacy of minimally invasive techniques in addressing complex congenital vascular disorders in children. This case adds further support to the safety and effectiveness of such interventions within the pediatric demographic, offering a viable option for patients who might otherwise encounter considerable risks associated with open surgical interventions. This research presents significant novelty across several critical dimensions. Firstly, it documents the inaugural successful closure of a pulmonary arteriovenous malformation (PAVM) in a pediatric patient utilizing the Lifetech® Konar-MF device, a procedure that has not been previously recorded in the existing medical literature. The transcatheter technique employed demonstrates effective outcomes with minimal complications when compared to traditional surgical interventions, thereby offering a safer and more practical alternative, particularly for younger patients. Furthermore, the findings indicate substantial clinical improvements in the subjects, including enhanced oxygen saturation levels and alleviation of symptoms such as dyspnea, cyanosis, and fatigue, which collectively contribute to a marked enhancement in the patients' overall quality of life. Lastly, the application of the Lifetech® Konar-MF device for embolization represents a significant advancement in medical technology, broadening the array of therapeutic options available for PAVM treatment. This integration of technological progress and clinical methodology underscores the innovative significance of this research within the field of pediatric cardiology.

Conflict of Interest: Each author declares that he or she has no commercial associations (e.g., consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

Authors Contribution: SFL (Concept, Design, Sources, Materials, Data Collection, Analysis and Interpretation, Literature Search), MAR and IKAU (Concept, Design, Analysis and Interpretation, Literature Search, Critical Review, TH (Concept, Design, Sources, Materials, Data Collection, Analysis and Interpretation, Literature Search). All authors approved the version to be published and agreed for all aspects of the article in ensuring that questions related to the accuracy or integrity of any part of the paper are appropriately investigated and resolved.

Declaration on the Use of AI: None.

Funding: The study did not receive any additional funding.

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Received: 4 December 2024
Accepted: 10 February 2025
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