

# Novel Technique in the Management of a Patient With Pulmonary Arteriovenous Malformation and Biventricular Non Compaction

**Desabandhu Vinayakumar**

Government Medical College Kozhikode

**Gopalan Nair Rajesh**

Government Medical College Kozhikode

**Haridasan Vellani**

Government Medical College Kozhikode

**Jomy Vadasseril Jose** (✉ [jomy960@gmail.com](mailto:jomy960@gmail.com))

Government Medical College Kozhikode <https://orcid.org/0000-0002-0369-4701>

---

## Research Article

**Keywords:** Pulmonary arteriovenous malformation, right to left shunt, device closure

**Posted Date:** June 2nd, 2021

**DOI:** <https://doi.org/10.21203/rs.3.rs-459335/v1>

**License:**  This work is licensed under a Creative Commons Attribution 4.0 International License.

[Read Full License](#)

---

# Abstract

**Background:** Transcatheter device closure has become the primary treatment modality for pulmonary arteriovenous malformations. When compared to lobectomy or vessel ligation, this approach is much less destructive and allows for selective closure of the arteriovenous malformation while preserving the normal branching vessels and lung parenchyma. But, often the procedure is cumbersome due to anatomical and procedural factors.

**Case Presentation:** We report the case of a 19 year old lady with biventricular non compaction and pulmonary arteriovenous fistula. She had deep cyanosis due to right to left shunt at two levels, through the stretched open patent foramen ovale due to severe right ventricular dysfunction and through the pulmonary arteriovenous malformation. The two feeders of the malformation were closed by device closure. The second feeder was closed by creating a venovenous loop via the patent foramen ovale, the pulmonary vein and through the pulmonary arteriovenous malformation into the pulmonary artery. To the best of our knowledge, this technique has never been described before in literature.

**Conclusion:** This is a novel technique in the closure of pulmonary arteriovenous malformations, particularly in tortuous vessels which augment the difficulty in the placement of the delivery sheath and delivery of the occluder device.

## Background

Transcatheter device closure has become the primary treatment modality for pulmonary arteriovenous malformations. When compared to lobectomy or vessel ligation, this approach is much less destructive and allows for selective closure of the arteriovenous malformation while preserving the normal branching vessels and lung parenchyma. But, often the procedure is cumbersome due to anatomical and procedural factors. We describe a novel technique in the closure of pulmonary arteriovenous malformations, particularly in tortuous vessels which augment the difficulty in the placement of the delivery sheath and delivery of the occluder device. To the best of our knowledge, this technique has never been described before in literature.

## Case Presentation

A 19 year old tribal girl presented with history of progressive effort dyspnoea of five years duration with bluish discoloration of nails and lips for the past three years. By means of clinical examination and multimodality imaging which included transthoracic echocardiography, cardiac CT scan, cardiac MRI scan and invasive cath study, she was diagnosed to have a biventricular non compaction with heart failure and a pulmonary arteriovenous malformation. She had deep cyanosis due to right to left shunt at two levels, through the stretched open PFO due to severe RV dysfunction and through the pulmonary arteriovenous malformation.

The case was discussed in the heart team meeting. Device closure of AVM to observe the improvement in saturation was the only feasible option as surgical lobectomy followed by a bidirectional Glenn was not approved as the LVEF was only 35%. We decided for selective closure of the two feeders, as a common trunk which gives rise to the two feeders could not be identified.

Device closure of one feeder of pulmonary AVM was done initially with a Amplatzer™ Vascular Plug II (AVP II) (Abbott Cardiovascular, Plymouth, Minnesota, USA) resulting in improvement in saturation to 88% from baseline value of 72% (Figure 1). But, repeated attempts for introducing sheaths or 5F Judkins Right (JR) 3.5 guiding catheter (Cordis, Santa Clara, California) into the other feeder from pulmonary arterial side was futile due to acute angulation at the origin of the second feeder. Stiff wires could not be introduced deep inside the feeder. Even a 4F catheter could not be negotiated deep into the feeder of AVM over exchange length coronary wires. An innovative technique of veno venous loop creation was done for introducing the guiding catheter into the other feeder. Using a 5F JR 3.5 guiding catheter a curved tip Terumo wire (Terumo Medical Corporation, Somerset, New Jersey) was introduced through inferior vena cava into right atrium and through patent foramen ovale into left atrium. The wire was then carefully introduced through the left lower pulmonary vein into the AVM and was manipulated through the AVM into the arterial feeder which was difficult to enter during previous attempts. By careful manipulations the wire could be advanced into the right pulmonary artery which was then snared out through the other femoral vein creating the veno venous loop (Figure 2,3). ( Wire through right femoral vein -> inferior venacava -> RA -> PFO -> left atrium -> pulmonary vein -> AVM -> arterial feeder -> pulmonary artery -> snared out -> RV -> RA -> inferior vena cava -> Left Femoral vein ). After loop creation the guiding catheter could be negotiated into the arterial feeder. A second AVP II device was deployed (Figure 4). After device closure the saturation improved on table to 92%. Patient was discharged with guideline directed medical management for heart failure. On follow up patient reported significant symptomatic improvement despite persistence of mild cyanosis. Repeat contrast echo showed persistence of right to left shunt at the level of ASD/PFO. There was mild improvement in LV systolic function with persisting restrictive RV filling pattern.

## Discussion

Transcatheter closure has become the primary treatment modality for pulmonary AVMs. Compared to lobectomy or vessel ligation this approach is much less destructive and allows for selective closure of the AVM while preserving the normal branching vessels and lung parenchyma. The AVP II is a cylindrical self-expanding device, made of nitinol wire mesh. The advantages of vascular plugs include easy release, reduced risk of migration and complete occlusion with a single plug.<sup>1</sup> But there are multiple challenges in achieving this feat. First of all, while attempting to deploy an Amplatzer type device which requires a large bore sheath, the support given by the guide wire is often inadequate. The problem is further complicated when there is vessel tortuosity. Beck et al used balloon-tipped catheters wedged in the distal vasculature and stiff guidewires to provide the support required to advance a device delivery sheath coaxially over it into the pulmonary AVM, and utilized braided sheaths to avoid sheath kinking.<sup>2</sup> Joseph et al describes a

technique wherein an atrial septal puncture was performed and a femoro femoral arteriovenous guidewire loop through the right pulmonary artery, pulmonary AVM, and left atrium was created. Traction on both ends of the guidewire loop allowed advancement of the device delivery sheath into the pulmonary AVM and successful completion of the procedure.<sup>3</sup> In our technique, the patient already had a stretched open PFO. But unlike in the case described by Joseph et al, we negotiated the guidewire in a retrograde fashion in to the pulmonary vein via the AVM and was snared out from the pulmonary artery. This technique provides additional support for the large delivery sheath and kinking of the device delivery sheath is unlikely to occur. Wire loop induced vascular injury may occur because of the traction on the vascular structure by the wire. This can be minimized by advancing the device delivery sheath and the Mullins sheath as far in as possible to reduce exposure of “bare” wire to vascular tissues.<sup>3</sup>

## Conclusion

We describe a novel technique in the closure of pulmonary arteriovenous malformations, particularly in tortuous vessels which augment the difficulty in the placement of the delivery sheath and delivery of the occluder device.

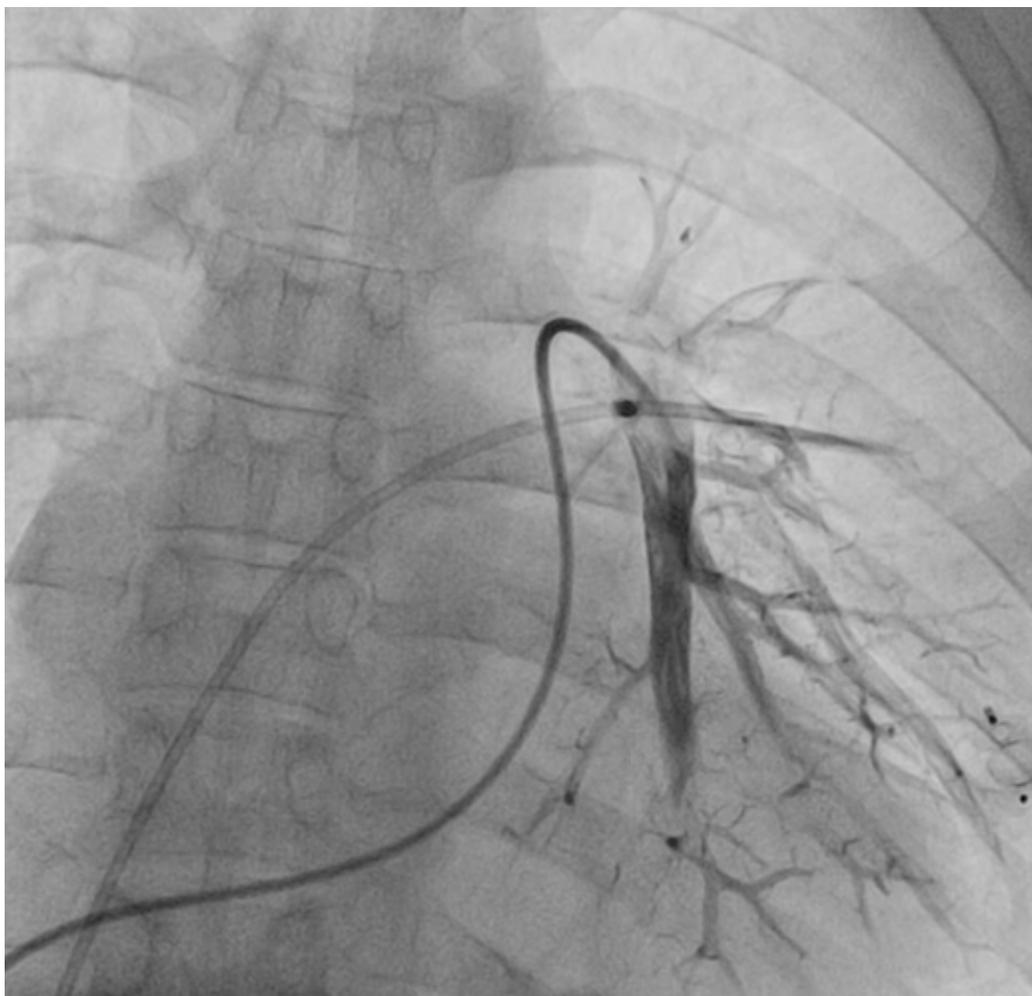
## Declarations

- Ethics approval and consent to participate - Obtained
- Consent for publication - Written informed consent was obtained from the patient for publication of this case report and any accompanying images
- Availability of data and materials – Obtained from Department of Cardiology, Calicut Medical College, Calicut, Kerala, India
- Competing interests – No competing interests
- Funding – No funding received
- Authors' contributions – DV and GNR was actively involved in performing the procedure described in the case report. JVJ was involved in the collection of data and materials. GNR and JVJ was involved in the preparation of the manuscript.
- Acknowledgements -Nil

## References

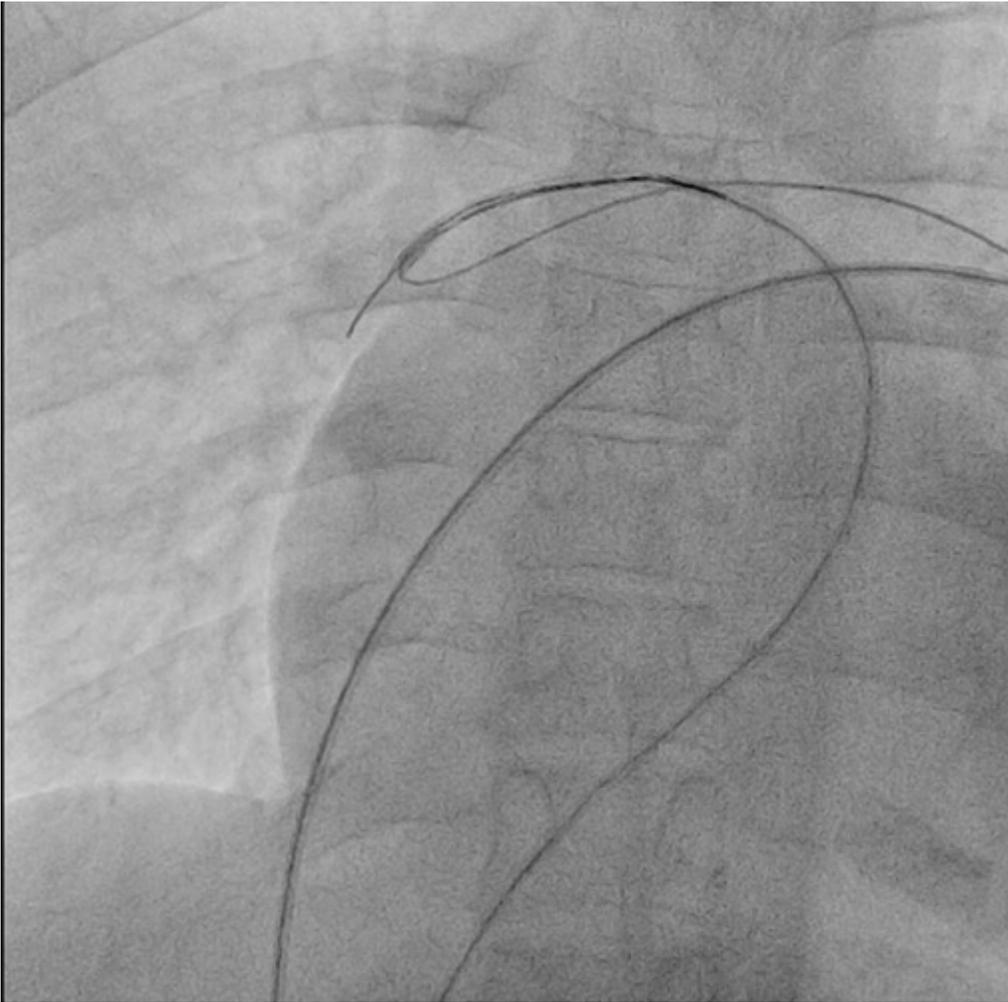
1. Rajesh GN, Sajeer K, Nair A, Haridasan V, Sajeer CG, Krishnan MN (2015) Device closure of pulmonary arteriovenous malformation using Amplatzer vascular plug II in hereditary hemorrhagic telangiectasia. *Indian Heart J* 67(5):455–458. doi:10.1016/j.ihj.2015.05.024
2. Beck A, Dagan T, Matitiau A, Bruckheimer E (2006) Transcatheter closure of pulmonary arteriovenous malformations with Amplatzer devices. *Cathet Cardiovasc Intervent* 67(6):932–937. doi:10.1002/ccd.20728

## Figures



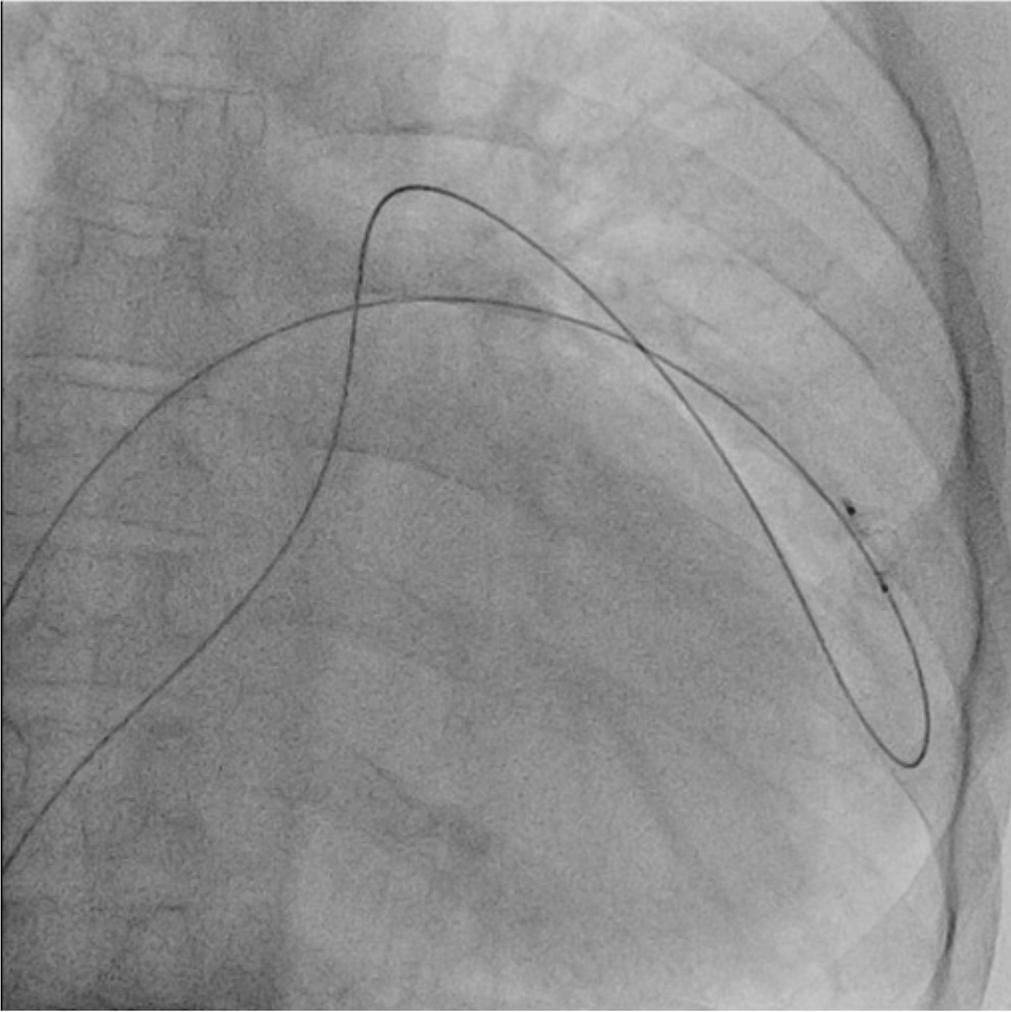
**Figure 1**

Selective left pulmonary artery angiography after the 1st device deployment showing the second feeder



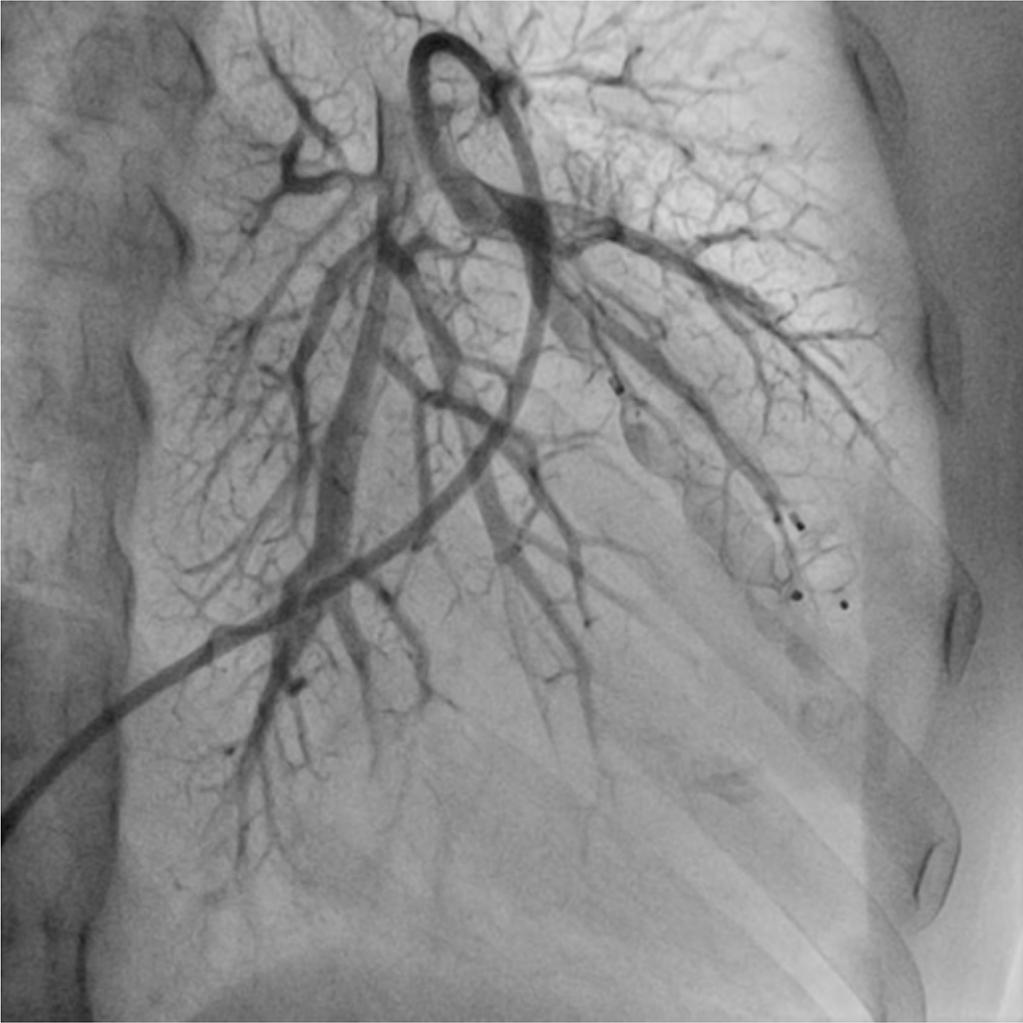
**Figure 2**

Snaring out the guidewire from the right pulmonary artery



**Figure 3**

The venovenous loop created



**Figure 4**

Complete closure of the two feeders by the two devices