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**Patent Foramen Ovale Combined with Pulmonary
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4 **TITLE PAGE**
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10 **Title of the manuscript:** Patent Foramen Ovale Combined with Pulmonary Arteriovenous
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15 **Short title:** PFO with PAVM
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21 **Conflict disclosure:** Jonathan Tobis serves as a consultant for W.L. Gore and Associates,
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23 Inc. and St. Jude Medical, Inc. All other authors have no financial conflicts of interest to
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27 disclose.
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Abstract

Both a patent foramen ovale (PFO) and a pulmonary arteriovenous malformation (PAVM) could be a source of a right-to-left shunt (RLS). A 52-year-old male who had a history of juvenile polyposis syndrome hereditary hemorrhagic telangiectasia (HHT) and a series of transient neurologic deficits, was found have multiple small PAVMs and a PFO. Percutaneous PFO closure was performed using a Gore Helex Septal Occluder, but the PAVMs were too small to close. A 71-year-old female without HHT who had a history of 2 strokes demonstrated a large RLS. Percutaneous PFO closure was performed using a GORE CARDIOFORM Septal Occluder and subsequent percutaneous PAVM closure using an Amplatzer Vascular Plug IV was done.

Text

INTRODUCTION

A pulmonary arteriovenous malformation (PAVM) is caused by an abnormal communication between a branch pulmonary artery and pulmonary vein, which occurs in about 1 in 2600 adults [1]. Approximately 70% of the cases of PAVM are associated with hereditary hemorrhagic telangiectasia (HHT) [2]. Conversely, approximately 15 to 35% of patients with HHT have a PAVM. Another etiology of a right-to-left shunt (RLS) is a patent foramen ovale (PFO), which is an inter-atrial communication present in about 20% of the general population [3]. In patients who have a PFO and a PAVM, it may be difficult to recognize that both entities co-exist.

CASE REPORT

Case 1: A 52-year-old male who had a history of juvenile polyposis syndrome-HHT with an occasional gastrointestinal tract bleed was referred to our institution for consideration of percutaneous PFO closure. The patient had a series of transient neurologic deficits and had a PFO documented by transesophageal echocardiography (TEE). Magnetic resonance imaging of the brain demonstrated lacunar infarctions. Chest computed tomography showed multiple small PAVMs which were considered to be too small to close. A right-heart catheterization was performed and bubble contrast intracardiac echocardiography (ICE)

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4 imaging showed a RLS. The PFO was closed using a 25-mm Gore Helex Septal Occluder
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7 (W.L. Gore and Associates, Inc., AZ, USA). After the device was deployed, bubble contrast
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10 ICE imaging did not show a residual RLS.

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12 Case 2: A 71-year-old female who had 2 strokes without significant atherosclerosis
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14 was referred to our institution for consideration of percutaneous PFO closure. A PFO with a
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16 large shunt was diagnosed by TEE. There was no family history of HHT and the patient had
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18 no telangiectasia or history of epistaxis. At right-heart catheterization, a J-tipped guidewire
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20 was unable to cross the inter-atrial septum. Angiography was performed to clarify the
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22 anatomy and confirmed the presence of a small PFO (Figure 1, Movie 1). Despite the small
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24 size of the PFO, in the presence of recurrent stroke, the PFO was closed using a 25-mm
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26 GORE CARDIOFORM Septal Occluder (W.L. Gore and Associates, Inc., AZ, USA). After
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28 device deployment, a persistent large RLS was demonstrated by bubble contrast ICE
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30 imaging. Selective angiography of the pulmonary artery demonstrated a PAVM with a feeding
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32 artery diameter of 6-7 mm (Figure 2A and 2B, Movie 2A). This was closed using an 8-mm
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34 Amplatzer Vascular Plug IV (St. Jude Medical, MN, USA) (Figure 2C, Movie 3).
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50 DISCUSSION

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52 The first case of a patient with HHT with multiple small PAVMs is unusual because a
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54 PFO was discovered as the cause of the RLS instead of a large PAVM. The current
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4 recommendations include embolization of all PAVM with a feeding artery of ≥ 2 mm if
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7 amenable for percutaneous closure [4]. However, there are no guidelines how to manage a
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10 concomitant PFO in the presence of a PAVM. Given that HHT patients have a high risk of
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13 bleeding, PFO closure to prevent recurrent cryptogenic embolism may be preferable to
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16 anti-platelet or anti-coagulable therapy.

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18 Our second case of a small PFO associated with a PAVM is unusual because the
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21 patient did not have a history consistent with HHT and thus a PAVM was not suspected. From
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24 our data base of a population who were referred for the assessment of PFO-related
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27 conditions, only 1 out of 806 subjects had a PAVM instead of a PFO in patients without HHT.
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30 In a separate data set at UCLA of 312 patients with HHT, there were 5 subjects who had a
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33 step up in RLS degree after release of the Valsalva maneuver suggesting a PFO in addition
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36 to a PAVM. Evaluation in 1 of these 5 demonstrated a PFO which was closed [5].

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38 The selective angiograms in our second case demonstrate a small PFO associated
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41 with a large PAVM. We suspect that the PAVM was the primary etiology of her strokes.
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44 However, the risk of paradoxical embolization is independent of the PAVM feeding artery
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47 diameter [6] and the size of a PFO is weakly associated with the incidence of cryptogenic
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50 stroke [7]. There are 3 reports describing cases with both a PFO and PAVM [8]–[10]. In all of
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53 these cases, the PFO was closed prior to the treatment of the PAVM.
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CONCLUSION

In patients who have a PAVM or PFO closed, and there is still a large residual RLS shunt, the presence of the other condition should be evaluated.

For Review Only

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3 **Figure legends**
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6 Figure 1: Right atrial angiography of a PFO
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9 Angiography shows the anatomy of a small PFO (arrowheads). The arrow shows the probe of
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11 intracardiac ultrasound catheter.
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15 Abbreviation: LA; left atrium; PFO, patent foramen ovale; RA, right atrium
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21 Figure 2: Pulmonary angiography of a PAVM
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24 A. A selective angiography of a PAVM (arrowheads). This shows an afferent branch
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26 pulmonary artery with a diameter of 6-7 mm (solid arrow) and an efferent branch
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28 pulmonary vein (dotted arrow).
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32 B. Three-dimensional angiographic reconstruction image of a PAVM. An afferent vessel
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34 (solid arrow) and efferent vessel (dotted arrow) are shown.
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38 C. An angiography after the embolization of a PAVM. Arrowheads show the position of
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40 the Amplatzer Vascular Plug IV.
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44 Abbreviation: PAVM, pulmonary arteriovenous malformation
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Supplementary Material Legends

Movie 1: Angiography of a patent foramen ovale

Movie 2: Angiography of a pulmonary arteriovenous malformation before the embolization

Movie 3: Angiography of a pulmonary arteriovenous malformation after the embolization

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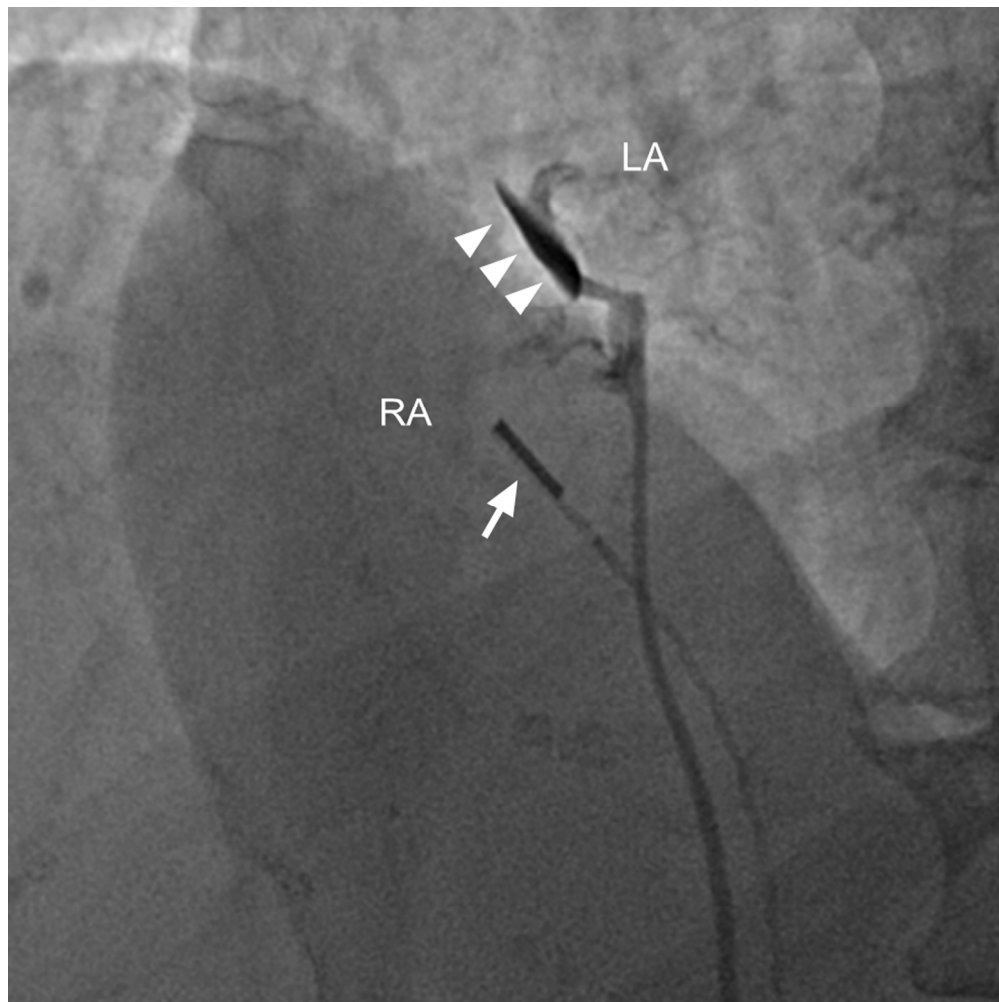


Figure 1: Right atrial angiography of a PFO
Angiography shows the anatomy of a small PFO (arrowheads). The arrow shows the probe of intracardiac ultrasound catheter.

Abbreviation: LA; left atrium; PFO, patent foramen ovale; RA, right atrium
166x165mm (300 x 300 DPI)

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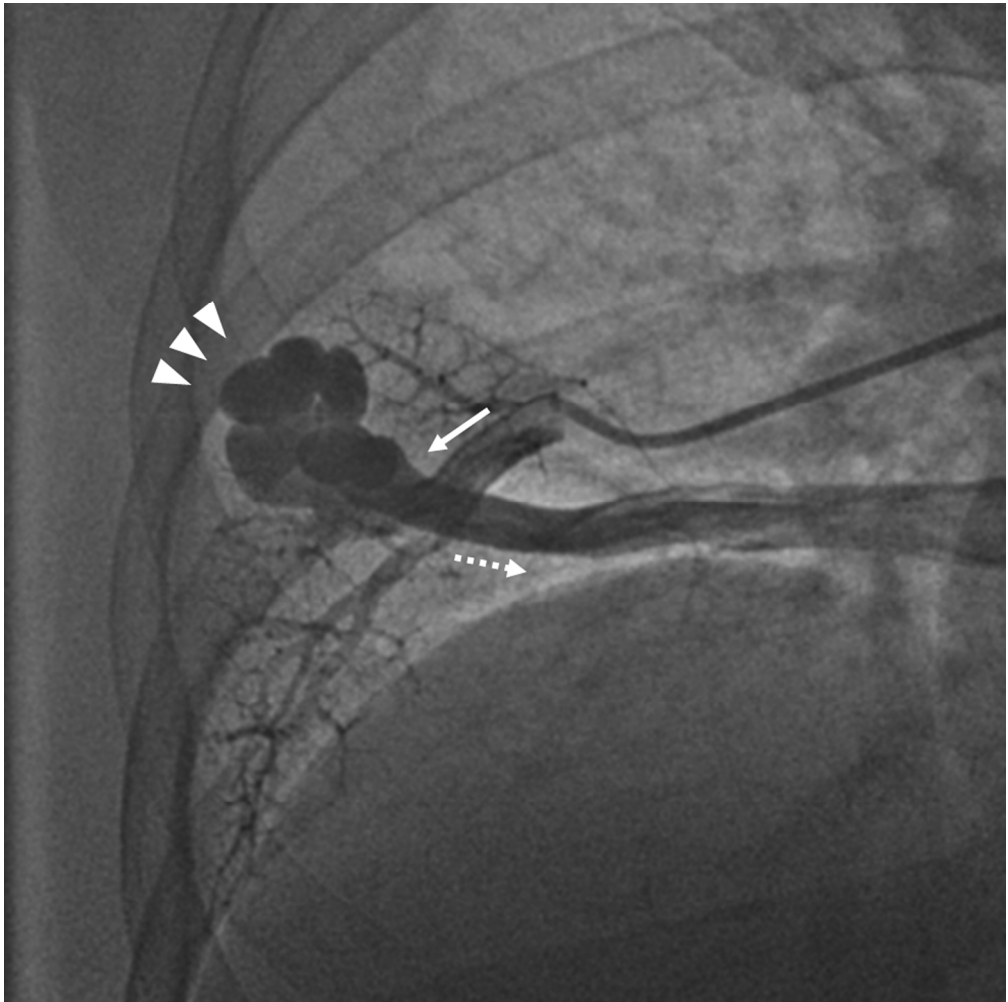


Figure 2A: Pulmonary angiography of a PAVM
A selective angiography of a PAVM (arrowheads). This shows an afferent branch pulmonary artery with a diameter of 6-7 mm (solid arrow) and an efferent branch pulmonary vein (dotted arrow).
166x165mm (300 x 300 DPI)

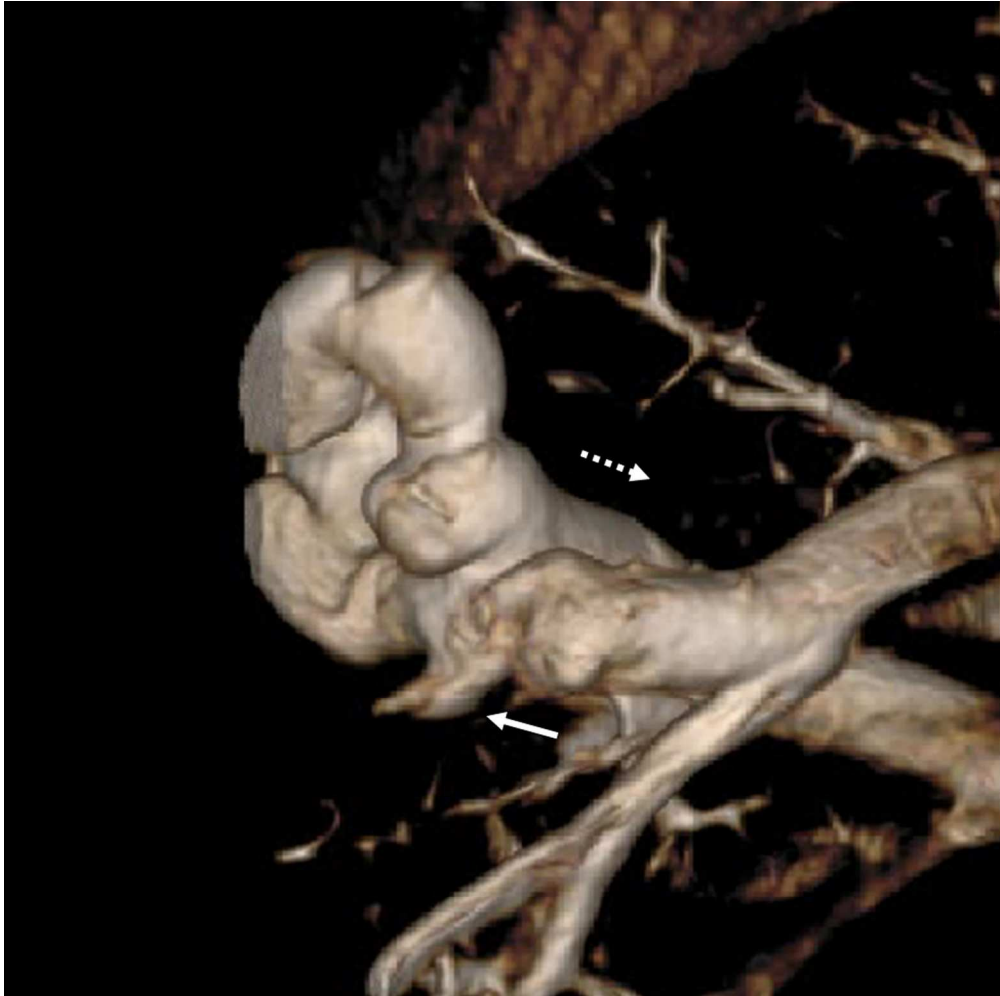


Figure 2B: Pulmonary angiography of a PAVM
Three-dimensional angiographic reconstruction image of a PAVM. An afferent vessel (solid arrow) and efferent vessel (dotted arrow) are shown.
166x165mm (300 x 300 DPI)

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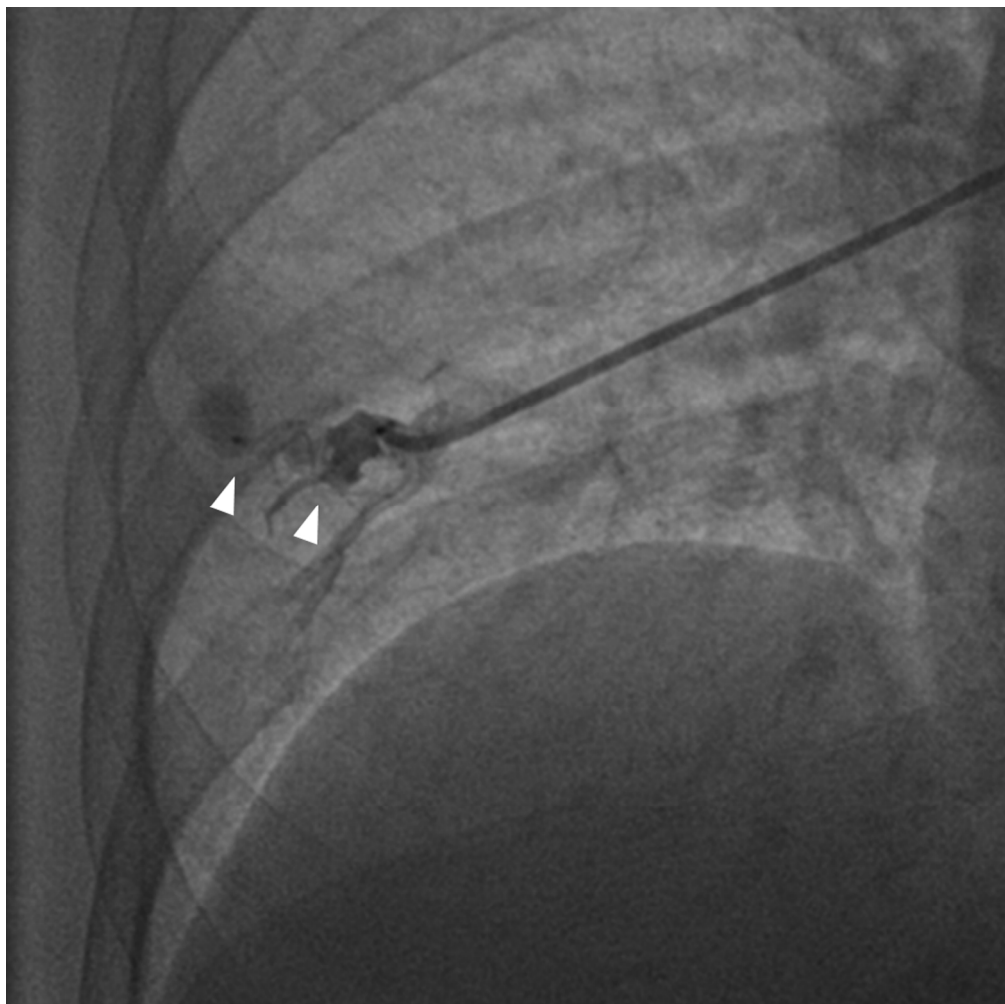


Figure 2C: Pulmonary angiography of a PAVM
C. An angiography after the embolization of a PAVM. Arrowheads show the position of the Amplatzer Vascular Plug IV.
Abbreviation: PAVM, pulmonary arteriovenous malformation
166x165mm (300 x 300 DPI)