



Vessel Embolization: Transcatheter Embolization of Pulmonary Arteriovenous Malformations and Aortopulmonary Collateral Arteries

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37.1 Transcatheter Embolization of Pulmonary Arteriovenous Malformations

37.1.1 Anatomic Description and Physiopathology

Pulmonary arteriovenous malformations (PAVMs) are direct high-flow, low-resistance fistulous communications between the pulmonary arteries and veins, bypassing the normal pulmonary capillary bed and resulting in an intrapulmonary right-to-left shunt.

Most PAVMs are congenital, with 80–95% cases are associated with hereditary hemorrhagic telangiectasia.

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Acquired PAVMs are even less frequent, occurring after heart surgery (Fontan or Glenn procedure), trauma, and pulmonary operations [1].

Most PAVMs are identified in the lower lobes, with the left lower lobe being the most common location. PAVMs are usually classified into single or multiple types. Approximately, 80% of PAVMs are simple in which the feeding arteries arise from one or more branches of a single segmental pulmonary artery.

The majorities of the rest are complex PAVMs, which have multiple segmental feeding arteries from more than one pulmonary segment. A smaller percentage of PAVMs are diffuse in which there is disseminated involvement of multiple pulmonary segments. Diffuse lesions are difficult to treat with transcatheter embolotherapy and are often referred for lung transplant.

PAVMs can be further characterized according to their radiological appearance. The fistula-type PAVM has a feeding artery directly connected to a draining vein, with a venous sac.

Less commonly, PAVMs are plexiform with a multiseptated aneurysm or a cluster of vascular channels [2].

37.1.2 Clinical Manifestation

Clinical manifestation of patients with PAVM varies depending on the size, number, and flow through the PAVM.

Most PAVMs, especially if they are small ones, can be clinically silent for a long time. However, large or diffuse malformations can cause a wide spectrum of clinical manifestations including exertional dyspnea, fatigability, cyanosis, and neurologic disorders (infarcts, transient ischemic attack, and brain abscesses) secondary to paradoxical embolism as well as life-threatening hemoptysis due to sac rupture.

Because the pulmonary capillary bed is bypassed, the blood flowing through a PAVM is not oxygenated and directly drained into the pulmonary veins, resulting in systemic hypoxemia.

In addition, the absence of the normal filtering of the pulmonary capillary allows particulate matter (air bubbles or clots) to enter into the systemic circulation leading to serious neurologic complications.

37.1.3 Indications and Patient Selection

Transcatheter embolization of PAVMs is indicated for patients who have evidence of significant systemic hypoxemia or for patients at risk for or who have a documented history of a paradoxical embolic event and also for the prevention of pulmonary hemorrhage [1].

The current guidelines recommend transcatheter embolization of PAVMs for all symptomatic patients and for asymptomatic patients with discrete lesions with feeding arteries greater than 3 mm in diameter [3].

For patients with complex or diffuse PAVMs that cannot be safely or completely occluded, a reduction in the volume of right-to-left shunting by partial or staged transcatheter closure may also be indicated in order to reduce cyanosis and alleviate symptoms.

37.1.4 Treatment Options

Therapeutic options for PAVMs include transcatheter embolization with coils or Amplatzer devices and surgical excision.

Currently, transcatheter embolization has become the mainstay of treatment for PAVMs. Surgical resection is currently rarely necessary and reserved for patients who are not candidates for embolization (e.g., in patients with diffuse lesions) or when embolization fails or is unavailable.

The primary aim of embolotherapy is to eliminate or reduce the right-to-left shunting to relieve desaturation symptoms, prevent pulmonary hemorrhage, and, most importantly, prevent neurologic complications associated with paradoxical embolism.

37.1.5 Pre-procedural Imaging

37.1.5.1 Contrast-Enhanced Echocardiography

Contrast-enhanced echocardiography is useful in the assessment of PAVM since it helps to distinguish between intracardiac and extracardiac shunts. Intracardiac shunts are characterized by the

visualization of bubbles in the left heart chambers within 1–2 cardiac cycles after appearing in the right atrium. In patients with PAVMs, this event occurs after a delay of 3–8 cardiac cycles. This delay is the time needed to traverse the pulmonary capillary beds and shunt back through the pulmonary veins to the left atrium.

37.1.5.2 Chest Computed Tomography

Multidetector CT (MDCT) has been established as the primary imaging modality in the detection of PAVM. CT angiography, especially with three-dimensional reconstruction, can provide important details to inform subsequent catheter-based treatment including the location, number, and size of the arterial feeding vessels and the presence of multiple, smaller malformations.

37.1.5.3 Contrast-Enhanced Magnetic Resonance Angiography (MRA)

Contrast-enhanced magnetic resonance angiography (MRA) has high sensitivity and specificity and should be considered in young patients, where radiation exposure will be of greater concern. It is potentially able to provide precise information on the number, location, and complexity of PAVMs.

37.1.6 Technique (Step-by-Step)

37.1.6.1 Diagnostic Pulmonary Angiography

Following femoral venous access is obtained, weight-adjusted unfractionated heparin (100 U/Kg) is given intravenously.

Routine right heart catheterization is performed to assess the pulmonary artery pressure.

The initial diagnostic pulmonary angiogram is usually performed in the anteroposterior (AP) projection and ipsilateral 40° oblique (this projection places the heart over the injected lung and spreads the basal segments) using a 6-Fr pigtail catheter or other catheters.

Complete angiography in both lungs prior to any attempt at embolization is mandatory in order to identify all feeder vessels to a PAVM, their diameter, and length. This determines the occlusion strategy.

37.1.6.2 Occluding Materials

The choice of the occlusion device is primarily based on the anatomy morphology and size of the vessel as well as on the personal experience and preference of the interventionist. In general, PAVM with feeding artery diameters of less than 5 mm may be treated with coils, whereas those with diameters larger than 5 mm should be preferably treated with ADO or AVP [4].

Coils

Magnetic resonance-compatible steel or platinum pushable or detachable coils are used in the majority of cases. It is recommended to choose coils that are at least 20–30% larger than the vessel to be occluded. The main drawbacks of coil occlusion include the risk of embolization, the need for multiple coils, the potential for recanalization, and the resulting long procedure time without complete occlusion. Pictures regarding COOK coils are reported in the chapter on ductus arteriosus closure. Of note, the MicroPlex cosmos complex coils (MicroVention, Terumo, Japan) or Axium™ helical or 3D detachable coil system (ev3 Neurovascular, Irvine, CA, USA), which is primarily used for the embolization of intracranial aneurysms and arteriovenous malformations, can also be off-label used for occlusion of PAVMs. These kinds of coils as well as interlocking detachable coils (IDCs, Boston Scientific, MA, USA) are usually used for packing the venous sac.

ADO

Before the advent of AVP, the Amplatzer duct occluder (ADO) device is used for the occlusion of medium-sized to large PAVMs. Several reports in the literature have described successful transcatheter treatment of large PAVMs using the ADO device. However, their application is limited by the need for relatively large long sheaths or large guiding catheters. With the availability of various kinds of AVP, the ADO has been rarely used for PAVM occlusion. Pictures regarding ADO are reported in the chapter on ductus arteriosus closure.

AVP

The AVPs are particularly suitable for embolization of large high-flow feeding vessels. They are a woven nitinol wire cylinder that can be delivered via small catheters such as standard 5–8-Fr coronary guiding catheters (Fig. 37.1a, b). The recent developed AVP IV (Fig. 37.1c) can even be introduced through a diagnostic catheter. During embolization, at least a 30–50% oversizing of the device to the feeding vessel is recommended for the prevention of device migration and total occlusion. The only potential drawback to the AVP appears to be the relatively long length of the occluder that may limit its use if the target vessel is too short.

37.1.6.3 Techniques for Closing PAVM with Coils

To date, the most common approach to closing PAVM is embolization of the feeding artery using pushable fibered or detachable coils delivered via coaxial catheters.

Once a PAVM and its feeding arteries had been identified, selective catheterization of the target vessels is performed using a coaxial guide system with an outer 6-Fr 80 cm guide catheter and inner 5-Fr 100 cm end-hole coil delivery catheter (i.e., multipurpose catheter, Cook).

The added support provided by the guiding catheter prevents the inner-coil delivery catheter from backing out of the target vessel during embolization, thereby allowing the coils to be delivered more precisely and in a tighter mass.

The use of such a coaxial guide system also allows smaller coils to be positioned within a larger anchor or scaffold coil.

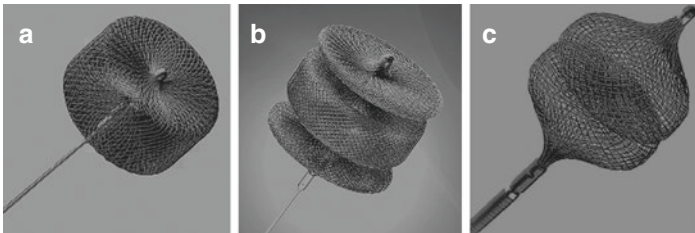


Fig. 37.1 Amplatzer plug I (a). Amplatzer plug II (b). Amplatzer plug IV (c)

Access to the middle and upper lobes can be challenging and is facilitated by the use of a 5-Fr Judkins left/right coronary catheter (cordis) or internal mammary catheter.

Once a feeding segmental artery is catheterized superselectively, the guiding catheter is advanced over the inner catheter to secure a stable position (i.e., placed in the parent segmental vessel), and the inner catheter is advanced into the vessel that feeds the malformation. The catheter tip is positioned beyond the origin of any proximal artery that supplies normal lung parenchyma.

Subsequently, hand-injected angiography (usually four frames) in multiple projections is performed to confirm position and define exact anatomy of the PAVM to determine site of implantation as well as size of the device to be used.

The basic principle for choosing the closure site is complete obliteration of the PAVM without compromising normal pulmonary parenchymal blood flow. Therefore, it is important to achieve as distal an embolization as possible in the feeding artery, preferably at the neck of the venous sac. This not only reduces the risk of occluding branches to normal adjacent lung, but may also reduce the likelihood of persistent perfusion of the venous sac by bronchial collaterals and of pulmonary artery recanalization. This is especially true when multiple PAVMs are present and multiple indiscriminate proximal occlusions could result in a significant reduction in pulmonary blood supply.

Coil size is an important consideration. Undersized coils are at risk to pass through the malformation and becoming an embolic agent, while oversized coils may be difficult to form a tight nest. Many interventionist empirically oversize the initial coil to the feeding vessel by at least 20%. After placement of the first coil, additional coils must be positioned until blood flow to the PAVM has ceased. In order to create a dense, cross-sectional occlusion for durable result, packing of subsequent smaller coils in the center of the first deployed coil is essential (Fig. 37.2).

The “anchor or side-branch technique” and “scaffold technique” have been documented to be very useful in achieving complete cross-sectional occlusion and avoiding paradoxical embolization of the coil via the PAVM [1].

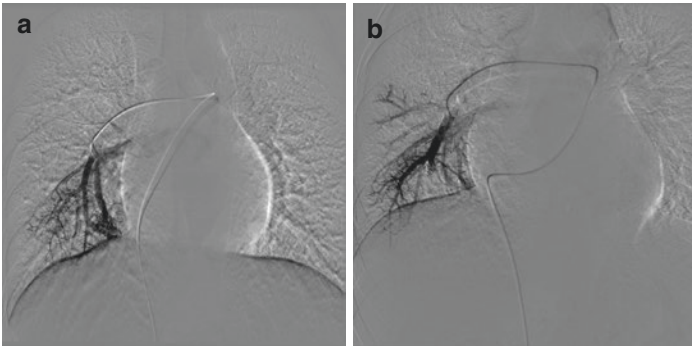


Fig. 37.2 (a) Pulmonary angiogram showing a PAVM of the right lower lobe. (b) Dense packing of two 6 mm COOK coils producing complete occlusion

The “anchor technique” is characterized by the first 2 cm of the coil and is purposely anchored in a side branch close to the aneurysmal sac, and the remainder of the coil positioned in the feeding artery and additional coils are densely packed so that cross-sectional occlusion is obtained. By securing the tip in a side branch, the risk of coil dislodgment is minimized.

The so-called scaffold technique is mainly used for high-flow vessels or when there is no anchoring vessel available. Initially, a high-radial force, fibered coil with a diameter 2 mm larger than the feeding artery is placed to create a scaffold. Then several small diameter high-radial force coils are placed as well into the endoskeleton, followed by several softer coils, until cross-sectional occlusion is obtained.

Packing of the aneurysm sac has been proposed as an alternative to feeding artery embolization when the feeding artery is too short to avoid sacrifice of large normal pulmonary artery branches or when the artery is a high-flow type with a higher risk of paradoxical embolization of coil.

Following the feeding artery that is catheterized superselectively with a 6-Fr guiding catheter, a coaxial microcatheter (i.e., a 2/2.6-Fr Excelsior microcatheter (Boston Scientific, MA, USA), 2.1/1.7-Fr Echelon™-10 microcatheter (ev3 Neurovascular, CA, USA), 2.4/1.7-Fr Headway microcatheter (MicroVention,

Terumo, Japan), etc.) is advanced coaxially through the catheter into the aneurysmal sac. After confirmation of the right position of microcatheter, venous sac embolization (VSE) with detachable coils can be performed. Several microcoils such as MicroPlex cosmos complex coils or Axium detachable coil system and IDCs are densely filled within the venous sac until a large matrix is established (Fig. 37.3). Sometimes, these detachable coils were

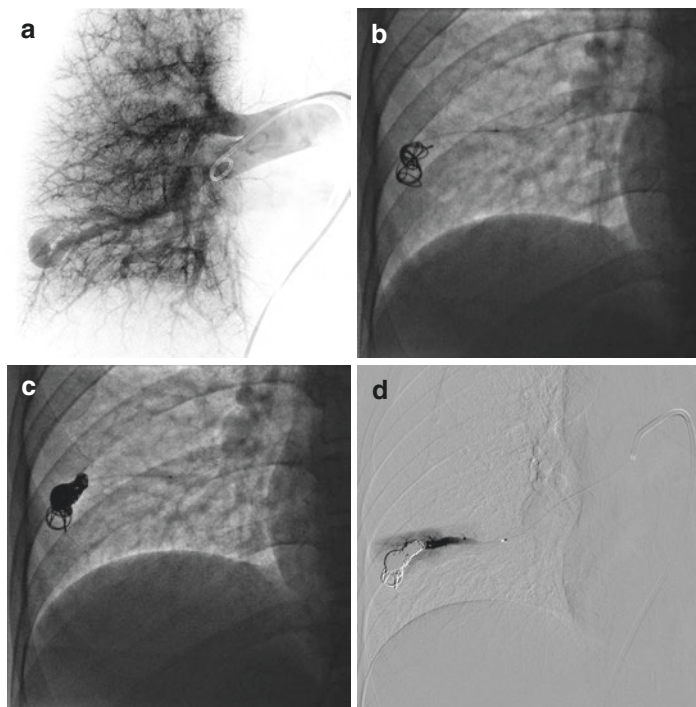


Fig. 37.3 A 49-year-old asymptomatic woman with simple-type PAVM treated by venous sac embolization. **(a)** Right pulmonary artery angiogram shows large basal PAVM of simple type with an aneurysmal venous sac. **(b)** Following an Echelon™-10 microcatheter placed in the aneurysmal sac, Axium™ helical detachable coils were sequentially deployed into the sac. **(c)** The sac was densely filled with 12 Axium helix coils, of which the largest one is 10 mm in maximum diameter. **(d)** Selective arteriogram a few minutes later demonstrates complete packing of the sac

used to form an initial wire mesh framework to prevent coil migration of the subsequent pushable small coils.

37.1.6.4 Techniques for Closing PAVM with Amplatzer Devices

For patients with PAVMs of large feeding artery or high-flow pattern, occlusion with coils is technically demanding and has a high risk of coil migration through the sac into the systemic circulation.

Alternatively, the recently developed Amplatzer vascular plug (AVP) appears to be an effective tool for embolization of PAVMs, particularly in patients with large outflow or short feeding arteries in whom embolization using coils entails a great risk of paradoxical embolization. At present, AVPs are the most commonly used devices for large PAVM occlusion.

The AVP is made from densely woven nitinol mesh wires that can be delivered via small catheters such as standard 5–8-Fr coronary guiding catheters and can be repositioned multiple times prior to its final release.

Following pulmonary artery pressure recording and angiography, the feeding artery is selectively cannulated using an appropriate-sized guiding catheter (5-Fr guiding catheter for AVPs 4–8 mm in diameter, 6-Fr for AVPs 10–12 mm in diameter, and 8-Fr for AVPs 14–16 mm in diameter).

Once a suitable position has been achieved as distally as possible within the feeding vessel and beyond any branches to normal lung, the AVP is then delivered to the target area.

The diameter of the AVP is selected to be at least 30–50% larger than the diameter of the feeding artery, according to the manufacturer's recommendation. Satisfactory positioning of the AVP is confirmed by repeat arteriography via the guiding catheter before its final detachment. If suboptimally positioned, the AVP is resheathed and redeployed in a more appropriate site. Since the AVP does not cause instantaneous thrombosis and in high-flow situations thrombosis typically takes up to 15 min, control angiography should be performed for at least 15 min after deployment of the occluder (Fig. 37.4).

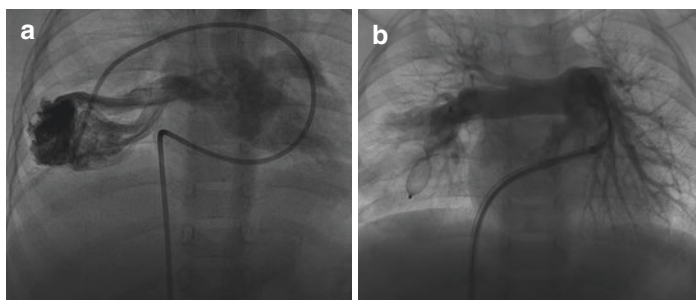


Fig. 37.4 Occlusion of a large right lower lobe PAVM with an AVP II. **(a)** Selective right lower lobe pulmonary angiogram demonstrates a large PAVM, which had a feeding vessel measuring 9 mm in diameter. **(b)** Control angiogram 10 min later after releasing a 14 mm diameter, AVP II demonstrates its optimal positioning at the neck of the PAVM and completes vessel occlusion with preservation of normal pulmonary artery branches. (This figure is provided courtesy of Prof. Yu-mei Xie, Department of Pediatric Cardiology, Guangdong Provincial People's Hospital, China)

The ADO can also be an alternative for closing large PAVM (Figs. 37.5 and 37.6). After catheterization of the feeding vessel, a long delivery sheath is introduced over a stiff exchange wire and placed in the feeding artery as close to the malformation as possible. The size of the ADO selected for embolization should be 2–4 mm larger than the caliber of the feeding vessel at site of implantation.

37.1.6.5 Techniques for Closing PAVM Using Venous Sac Embolization (VSE) in Combination with Feeding Vessel Embolization (FVE)

The combined treatment approach including the VSE using the detachable coils, followed by occlusion of the large feeding arteries using the AVP devices, has also been documented to be a highly efficient method for the treatment of the complex PAVMs with large outflow vessels or short feeding arteries. The combination of these two occlusion techniques enhances the ability of clot formation in the PAVM. The decrease of blood inflow into the aneurysmal sac due to the application of the AVP in the feeding

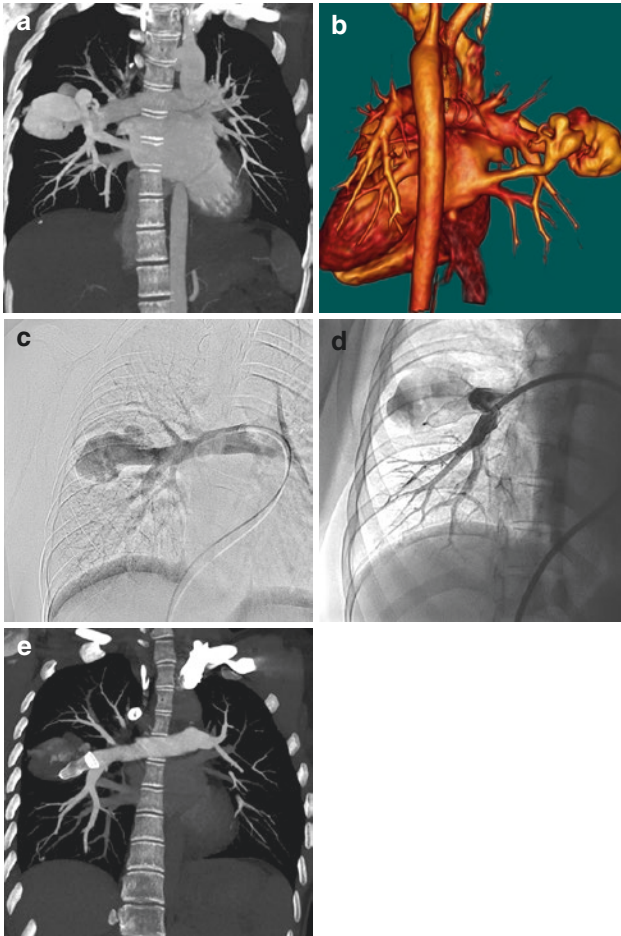


Fig. 37.5 A female patient had embolization of a right middle lobe PAVM with an ADO. (a) Initial three-dimensional reconstructed CT scan of the chest and (b) 3D volume-rendered CTA images showing a very large PAVM in the right middle lobe. (c) Right pulmonary artery angiogram demonstrating a large PAVM, with a feeding vessel measuring approximately 12 mm in diameter at its point of communication with the aneurysmal venous sac. (d) Angiogram after positioning a 16–18 mm ADO demonstrates its optimal positioning at the neck of the PAVM, and the PAVM was successfully occluded. (e) A follow-up CTA 1 month after embolization demonstrates the PAVM remains occluded. (This figure is provided courtesy of Prof. Yuan Feng, Department of Cardiology, West China Hospital, China)

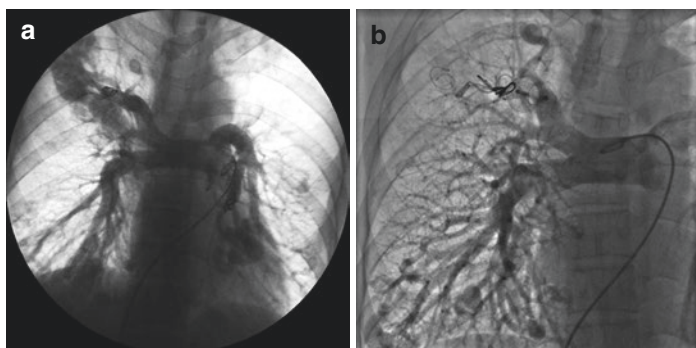


Fig. 37.6 A 29-year-old man with multiple PAVMs was referred for repeated PAVM occlusion. (a) Pulmonary angiogram revealed recanalization of previously embolized PAVMs. (b) PAVMs in the right upper and lower lobe were completely occluded with ADO

vessels increases the blood clotting ability and improves the occlusion of the aneurysmal sac. The steps of dense packing of venous sac using detachable coils combined with FVE using the AVP devices have been described above (Figs. 37.7 and 37.8).

After embolization of the feeding artery or arteries by any of the above methods, repeated segmental and lobar angiography should be performed to assess for complete occlusion and any accessory feeding vessels that may have been missed on the initial planning run and might also require embolization.

37.1.7 Expected Results

1. Pulmonary angiogram postembolization confirmed complete occlusion of the PAVMs.
2. A significant improvement in systemic arterial oxygen saturation and sustained relief of clinical symptoms attributed to the PAVMs on follow-up are obtained after embolization.
3. Contrast-enhanced CT at follow-up showed that the PAVMs remained occluded, with a significant shrinkage of the vein sac or complete resolution of the malformations.

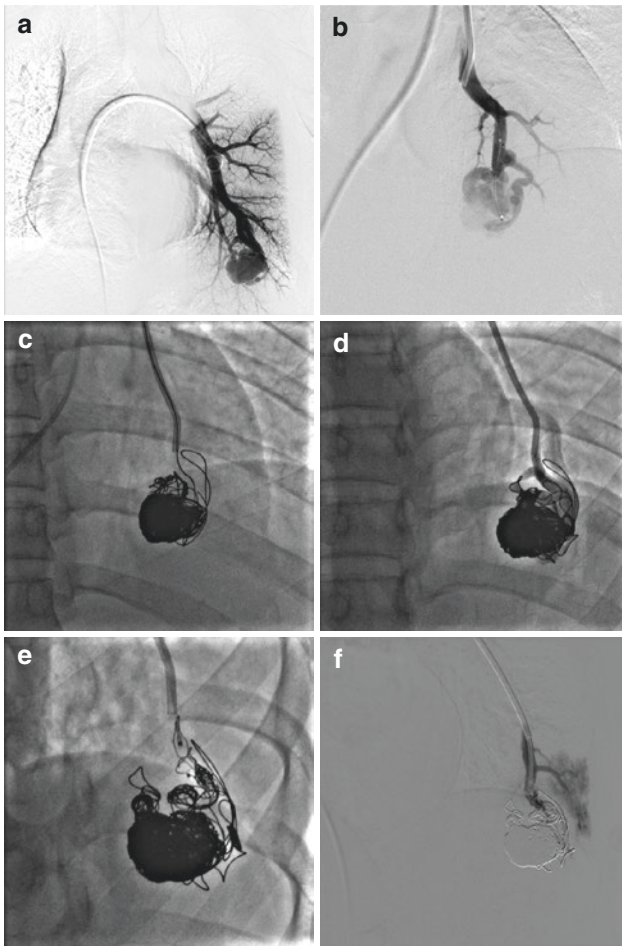


Fig. 37.7 A 25-year-old woman with hereditary hemorrhagic telangiectasia (HHT), large PAVM, and history of transient ischemic attack treated by VSE combined with FVE. **(a)** Left pulmonary arteriogram shows a complex-type PAVM with three feeding artery and a venous sac. **(b)** A 2.4/1.7 Fr Headway microcatheter was advanced coaxially through the 6-Fr MPD guiding catheter into the sac. **(c)** Twelve MicroPlex cosmos coils are densely filled within the venous sac. **(d)** There is still flow through the malformation following coil embolization of the aneurysmal sac. **(e)** A 6 mm AVP I was deployed at the neck of the PAVM. **(f)** Selective arteriogram a few minutes later demonstrates complete vessel occlusion with preservation of normal proximal pulmonary artery branches

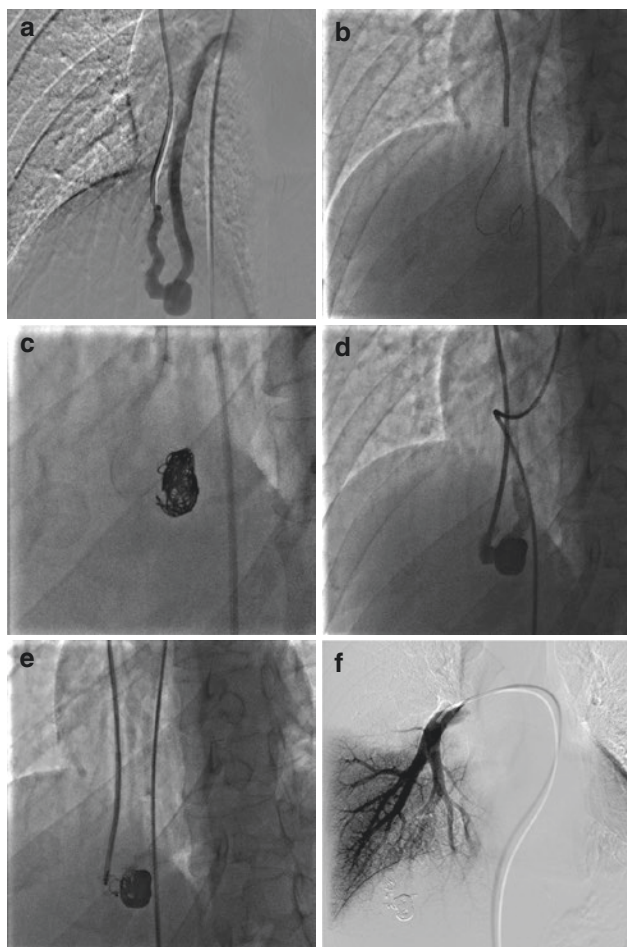


Fig. 37.8 A 35-year-old man with large bilateral PAVMs, and history of surgical resection of left lower lobe PAVM, was treated by VSE combined with FVE. (a) Subselective right pulmonary angiogram demonstrates a simple-type large PAVM with 7 mm feeding vessel, an aneurysmal venous sac. (b) A coaxial 2.4/1.7 Fr Headway microcatheter was placed in the aneurysmal sac. (c) The venous sac was densely packed with eight MicroPlex cosmos coils and one MWCE-35-5/6 mm coil. (d) Control angiogram performed 5 min later showing significant residual shunt. (e) A 10 mm AVP I was deployed at the neck of the PAVM. (f) Angiogram performed a few minutes later demonstrates complete vessel occlusion

37.1.8 Complications and Its Management

To date, there is no mortality occurred during the procedure and long-term follow-up. The complications of transcatheter embolization of PAVMs documented in the literature have been infrequent and are listed as follows.

37.1.8.1 Device Embolization

Device migration with paradoxical embolization is one of the most severe complications and occurs in 0.7–3% of treated patients, especially in cases of high-flow malformations with large outflow vessels. Paradoxical coil embolism into the cerebral artery, left popliteal artery, and left carotid artery has been reported. The choice of appropriate-sized coils is crucial to minimize the risk of coil embolization. The “anchoring” and “scaffolding” techniques are also frequently applied to overcome the problem of coil migration. In PAVMs patients with big-sized feeding arteries (>5 mm), coils probably should not be the first choice for embolization, and AVP alone or in combination with coils might be a better primary option for embolization in these patients.

37.1.8.2 Pulmonary Infarction

Pulmonary infarction has been observed in about 3% of patients. It usually occurs when the embolization causes occlusion of normal branches secondary to overly proximal positioning of embolization materials. To minimize this event, the embolization materials should be placed as close to the PAVM and as distal to normal side branches as possible.

37.1.8.3 Air Embolization

Air embolization is not infrequently encountered during the procedure. This usually occurs when a catheter or wire is withdrawn rapidly out of the sheath. When blood cannot replace the space previously occupied by the retrieved catheter, air will be sucked into the delivery sheath. Air accidentally enters into the coronary arteries causing acute chest pain, bradycardia, and temporary ECG

changes. This usually resolves within 15min. A continuous flushing of catheters, observation for back-bleeding, and removal of catheters or wires “underwater” can largely prevent this complication.

37.1.8.4 Pleurisy

Pleurisy is the most frequent complication of embolization occurring in approximately 15–31% of patients. Delayed pleurisy (4–6 weeks after the procedure) with fever and infiltrates has been reported mainly with larger PAVMs. It is thought that this is due to delayed thrombosis of the aneurysmal sac and is usually self-limited and responsive to nonsteroidal anti-inflammatory drugs.

37.1.8.5 PAVM Recurrence

Recurrence of PAVMs can occur in 15% of cases, but is not considered a real complication or failure of the treatment, since it can result from recanalization of previously occluded PAVMs, collateral reperfusion about the embolized site from adjacent small pulmonary branches that have grown or developed over time, collateral flow from bronchial or other systemic arteries distal to the occlusion site, or missed accessory pathways. Risk factors for recanalization have been established, including large feeder vessel, poor coil packing, use of an insufficient number of coils or coil elongation (use of oversized coils), complex PAVMs, and embolization more than 10 mm from the venous sac. This complication can be reduced by good closure technique and by selecting appropriate occluder for large PAVM.

37.1.9 Post-procedural Care and Follow-Up

Most patients can be discharged on the next day following the procedure. For patients with large feeding arteries and received large occluder, daily oral aspirin (5 mg/kg/day) is recommended for 6 months to prevent thromboembolic complications. Prophylactic antibiotics are not routinely recommended for all treated patients. In patients with incomplete occlusion with residual shunting, physicians should be aware of the risk of mechanical

hemolytic anemia. Care should also be taken to early detect femoral thrombosis and local hematoma at puncture site.

Long-term follow-up of treated patients with imaging modality and clinical and physiologic evaluation should be performed in order to document recanalization of embolized PAVMs early, as well as to detect growth or enlargement of the untreated small lesions. It is recommended that a combination of clinical evaluation, physiologic testing, and contrast-enhanced CT scan is the best algorithm of follow-up.

37.2 Transcatheter Embolization of Aortopulmonary Collateral Arteries

37.2.1 Anatomic Description and Physiopathology

Aortopulmonary collateral arteries (APCs) can be detected in association with various congenital heart diseases (CHDs), from simple malformations to complex cyanotic CHDs such as tetralogy of Fallot, pulmonary atresia, and single ventricle with pulmonary stenosis, resulting in varying degrees of left-to-right shunting.

APCs may be masked by another predominant cardiac lesion and are not discovered until after surgical repair of the major lesions. They are often found in cyanotic CHDs patients who have undergone a Glenn shunt procedure or Fontan repair.

The APCs typically originate from the anterior wall of the descending thoracic aorta at the level of the carina. However, they can also arise from the lower descending thoracic and abdominal aorta or innominate arteries.

They frequently run a retroesophageal course. Occasionally, the collateral arteries may arise from the coronary artery.

In patients with cyanotic CHD and reduced pulmonary blood flow, the additional pulmonary blood flow provided by APCs can relieve systemic hypoxemia prior to surgical correction. However, APCs' flow can result in significant volume overloading of the left ventricle, compete with and limit blood flow via the pulmonary arteries, and increase pulmonary arterial pressure and vascular resistance during the postoperative period of corrective surgery.

37.2.2 Clinical Manifestation

Small APCs are usually clinically silent, but large or multiple APCs can result in pulmonary overperfusion and symptomatic cardiac volume overload manifested as exertional dyspnea, recurrent pleural effusion, protein-losing enteropathy, frequent lower airway infection, and hemoptysis.

37.2.3 Indications and Patient Selection

37.2.3.1 Indications

The decision to occlude APCs primarily depend on the degree of left-to-right shunting and degree of dual supply between the aortopulmonary collateral and the native pulmonary artery to the segmental branches and pulmonary vascular bed. Transcatheter occlusion of APCs is indicated for the treatment of aortopulmonary collateral vessels with documented large left-to-right shunting in biventricular or single-ventricle physiology that results in congestive heart failure, pulmonary overcirculation, and respiratory compromise, or development of pleural effusion or protein-losing enteropathy [3].

37.2.3.2 Relative Indications

1. Transcatheter occlusion of APCs may be considered in the presence of moderate-sized collaterals found in asymptomatic single-ventricle patients undergoing routine pre-Glenn or pre-Fontan cardiac catheterization.
2. Transcatheter occlusion of APCs may be considered in patients with pulmonary atresia and aortopulmonary collaterals that have adequate dual supply from native pulmonary arteries.

37.2.3.3 Contraindications

1. Transcatheter occlusion is not recommended for the presence of APCs of any size in biventricle or single-ventricle patients who have significant cyanosis due to decreased pulmonary flow.

2. Transcatheter occlusion is not recommended for patients in whom the responsible collateral arteries directly supply a large area of pulmonary parenchyma, when embolization could result in infarction of the lung parenchyma [3].

37.2.4 Treatment Options

Therapeutic options for APCs include transcatheter embolization and surgical ligation. Surgical ligation can be technically challenging because of the identification, and dissection of the APCs can be very difficult, especially when they are transdiaphragmatic, and the operative field can be flooding by APCs supplying. Transcatheter occlusion is currently the preferred method for the management of APCs. The primary goal of embolotherapy is to control excessive flow of blood to the lungs.

37.2.5 Pre-procedural Imaging

Conventional angiography remains the gold standard for morphological assessment of the APCs. Noninvasive imaging modalities such as contrast-enhanced MRA and multidetector-row computed tomography (MDCT) with three-dimensional reconstruction are also useful for the assessment of APCs. Both of them can clearly identify the number, origins, course, and diameter of the APCs.

37.2.6 Technique (Step by Step)

37.2.6.1 Aortography and Pulmonary Angiography

Access is obtained in both the femoral artery and vein. Systemic anticoagulation (heparin 100 U/Kg) is provided, and the activated clotting time is maintained within the therapeutic range. A standard right heart catheterization is performed to assess the degree of shunting and evaluate the pulmonary artery pressure. A diagnostic aortogram and pulmonary angiogram is performed with a 5-Fr pigtail catheter. The goal is to identify the anatomic

characteristics of the APCs including the number, origin, course, diameter, and flow distribution pattern of the APCs and the presence or absence of native pulmonary arterial supply in the region of the “target” vessel. Since there is considerable anatomic variation among APCs in their origins (it can arise anywhere along the aorta or its major side branches), course, and branching patterns, selective angiography at multiple sites is required to fully assess for APCs. In some circumstances, it may also be necessary to perform selective angiography of the right or left subclavian artery, and even the coronary arteries, to fully disclose the collateral arteries.

37.2.6.2 Occlusion Techniques and Devices

Currently, transcatheter occlusion of APCs is performed most commonly with detachable or undetachable coils and AVP. Device selection is made according to the angiographic features of the target vessels.

Following diagnostic aortogram, the target collateral arteries are selectively engaged using a coaxial guide system with an outer 6-Fr Judkins right guiding catheter (Cordis, USA) or Cobra catheter (Terumo, Japan) and inner 5-Fr multipurpose catheter (COOK Corp., USA). The use of coaxial catheters allows for deep coil delivery and reduces the risk of proximal coil malposition. Once a suitable position has been achieved as deeply as possible within the target vessel, the appropriate-sized coils are then delivered to the target vessel. If undetachable coils are used for occlusion, the target vessels are selectively catheterized with a 5-Fr Judkins right guide catheter or Cobra catheter through which a microcatheter is introduced. The use of a coaxial microcatheter avoids the risk that a catheter may be dislocated by tension during the advancement of microcoils and the subsequent problem of coil deployment in an inappropriate systemic artery. With desired catheter position obtained, microcoils are delivered into the target vessel by saline flush.

The AVP is particularly suited for embolization of large, short, high-flow, or tortuous collateral arteries, where coil migration is possible or multiple coils may be needed. An appropriate-sized guiding catheter or long sheath is advanced over a

hydrophilic-coated guidewire into the collateral artery as deeply as possible. The AVP is then advanced via the guiding catheter or sheath into the vessel. Hand-injected angiogram is performed 15 min later to confirm coils or AVP position within collateral arteries.

It is important to recognize that APCs may have multiple sources of arterial supply, and occlusion devices should be delivered as selectively and as deeply into the target vessel as possible to block all potential arterial supply to the final pulmonary exit point.

Herein, a few clinical cases are briefly presented to illustrate the transcatheter occlusion techniques:

Case 1 A 1-year-old boy had successfully undergone surgical correction for double-outlet right ventricle and transesophageal echocardiography-guided device closure of muscular ventricular septal defect. At surgery, there was no evidence of increased pulmonary venous return. On postoperative day 3, attempts to wean the child from mechanical ventilation after surgery were unsuccessful. The patient developed progressive congestive heart failure and need for escalating doses of inotropic support after surgery. He was referred for transcatheter occlusion of multiple APCs (Fig. 37.9).

Case 2 A 6-month-old boy who had undergone surgical correction for pulmonary atresia and repair of atrial septal defect and ventricular septal defect was referred for emergent transcatheter occlusion of APCs due to a progressive low-output syndrome and the need for escalating doses of inotropic support after operation (Fig. 37.10). Transcatheter embolization of the APC resulted in prompt improvement in patient's conditions and rapid weaning from mechanical ventilation support.

Case 3 A 4-month-old boy had undergone a successful surgical correction for T.O.F. However, the patient was failure to wean from mechanical ventilation support. The postoperative period was further complicated by airway infections and dependence on inotropic support due to postoperative left ventricular dysfunc-

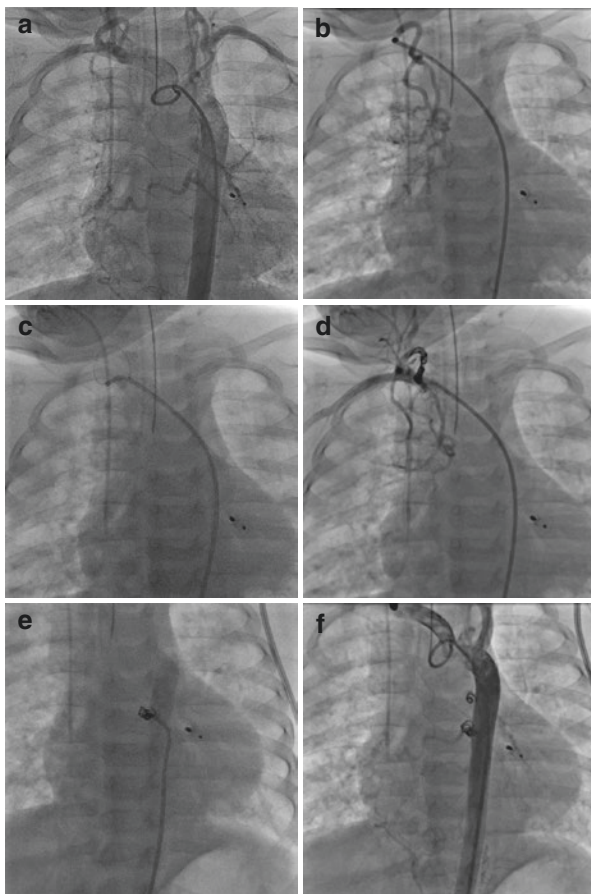


Fig. 37.9 (a) Ascending aortogram showing multiple APCs arising from the right subclavian artery and descending aorta. (b) The APC artery arising from right subclavian artery was selectively engaged by a 5-Fr Cobra catheter (Terumo, Tokyo, Japan) through which a 0.014 in. guidewire was advanced into the target vessel distally. (c) Over the guidewire, a 1.9-Fr, 130 cm Instant-pass microcatheter (APT Medical, Hunan, China) was introduced into the target vessel as deep as possible. (d) Subsequently, three MWCE-18S-5/2 mm tornado coils (COOK Medical, IN, USA) were advanced via the microcatheter by the 0.014 in. guidewire into the target vessel. The coils were packed densely at its narrowest segment. Following placement of coils, selective right subclavian artery angiogram demonstrates blood flow of the collateral artery was significantly decreased. (e, f) The remaining two APCs arising from the descending aorta were successfully treated in the similar fashion with two and one MWCE-18S-5/2 mm tornado coil, respectively

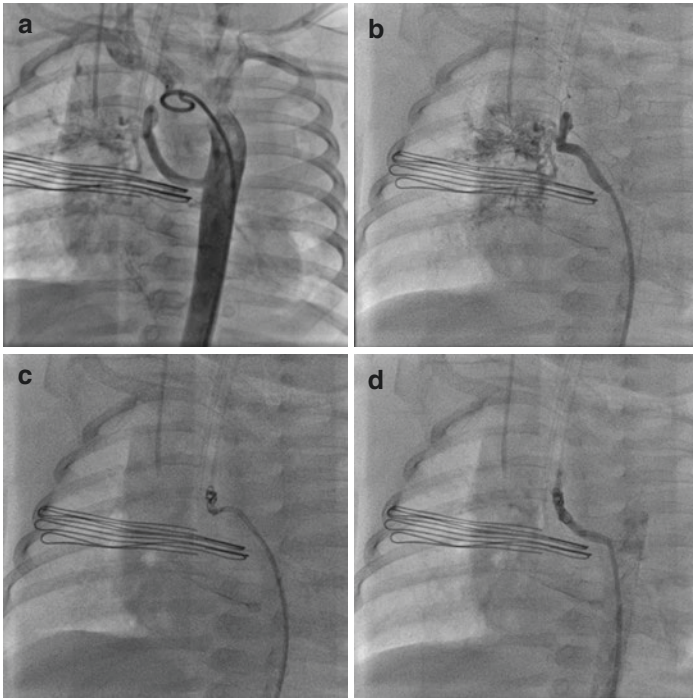


Fig. 37.10 (a) An angiogram in the aorta arch demonstrating a large APC artery arising from the descending aorta, which mainly supplying the right lung. (b) The APC artery was selectively engaged with a 5-Fr Judkins right diagnostic catheter (Cordis, FL, USA). The catheter was placed as distally as possible within the target vessel. (c) Two coils (MWCE-35-5/3 mm, MWCE-35-3/4 mm coils, COOK Medical, IN, USA) were sequentially advanced via the 0.035 in. hydrophilic wire (Terumo, Tokyo, Japan) and placed at the narrowest part of the collateral artery. (d) Following placement of coils, selective angiogram demonstrating satisfactory occlusion of this collateral artery

tion. He was referred for transcatheter occlusion of APCs (Fig. 37.11).

Case 4 A 2-month-old boy diagnosed with ventricular septal defect and pulmonary atresia was referred for transcatheter occlusion of APCs prior to surgical correction, to avoid excessive pul-

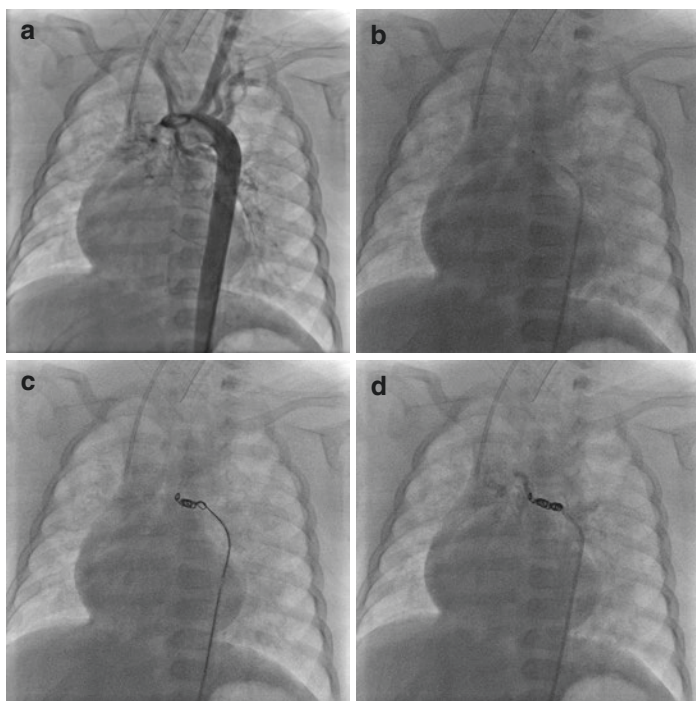


Fig. 37.11 (a) Angiogram in the aorta arch demonstrating a large APC artery arising from the descending aorta, which supplying bilateral lungs. (b) The target collateral artery was cannulated selectively using a coaxial guide system with an outer 5-Fr Cobra catheter (Terumo, Tokyo, Japan) and inner Instantpass microcatheter (APT Medical, Hunan, China). (c) Then, four tornado coils (MWCE-18S-4/2 mm, 18S-3/2 mm coils, COOK Medical, IN, USA) were sequentially advanced via the microcatheter by the 0.014 in. guidewire and placed at the narrowest segment of the target vessel with dense coil packing. (d) Selective angiogram after deployment of coils demonstrating complete occlusion of the collateral artery

monary blood return during the corrective surgery. Transcatheter embolization of a large APC using the AVP in combination with coils (Fig. 37.12). Considering the patient's condition could not tolerate long operation time, the planned procedure for occlusion of the remaining APC artery was aborted.

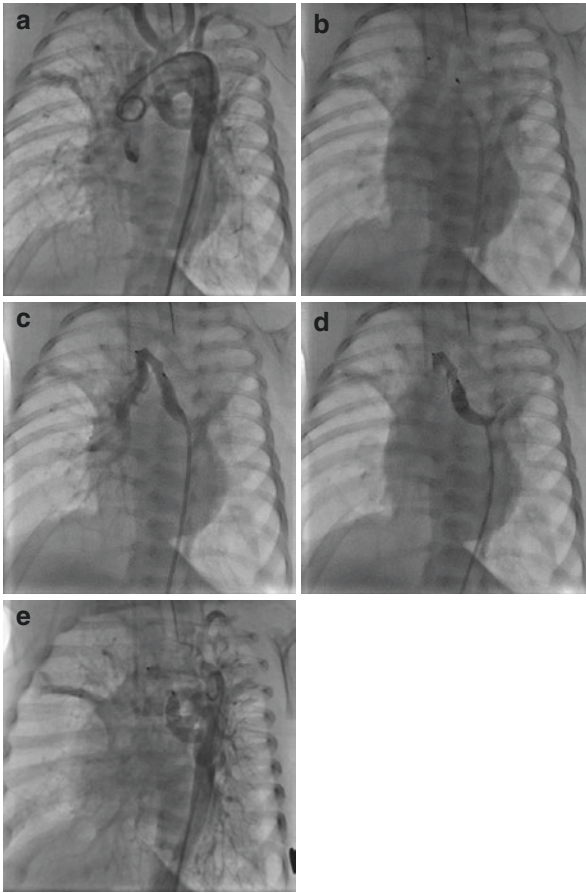


Fig. 37.12 (a) An angiogram in the ascending aorta demonstrating the presence of multiple APCs arising from the descending aorta, which supplying bilateral lungs. The larger one measuring approximately 4 mm in diameter at its narrowest. (b) The larger collateral vessel was selectively engaged with a 5-Fr Judkins right 4.0 guiding catheter (Cordis, FL, USA), and an 8 mm AVP (AGA Medical, MN, USA) was deployed at its narrowest position. (c) Control angiogram was performed 5 min later after the AVP deployment, demonstrating significant residual shunt. (d) Therefore, an addition MWCE-18S-8/5 mm Tornado coil (COOK Medical, IN, USA) was implanted, resulting in immediate, complete occlusion of this vessel. (e) Repeat descending aortogram performed 10 min later, demonstrating no residual shunting across this collateral vessel

37.2.7 Expected Results

1. Cardiac catheterization postembolization confirmed complete occlusion of the APCs with the pulmonary arterial pressure and oxygen saturation decreased, while the systemic pressure elevated to the normal level.
2. Significant improvement or resolution of symptoms attributed to the APCs at physician follow-up is obtained after embolization.

37.2.8 Complications and Its Management

37.2.8.1 Device Embolization

Device embolization into an important systemic artery occurs in about 1% of embolization attempts, mainly with coils. It usually occurs when a coil can't be fully accommodated by the target vessel that led to coil bouncing out of the collateral during or after implantation. In order to avoid such a complication, the selected coils should be of appropriate size, and it should be placed as deeply into the target vessel as possible. When coil embolization occurs, retrieval of the coil with a snare may be considered.

37.2.8.2 Pulmonary Infarction

The complication of pulmonary infarction occurs when the APCs constitute the sole supply to the affected lung or the responsible collateral arteries supply directly a large area of pulmonary parenchyma. Prior to embolization, a careful analysis has to be made based on the collateral circulation to ensure that the collateral arteries targeted for embolization are not the sole source of flow of blood to a parenchymal segment.

37.2.8.3 Hemolysis

Hemolysis has been rarely reported with embolization of APCs. This rare complication occurs if there is significant residual shunting across the occluder. Once it occurs, the patient should be monitored and treated medically. If hemolysis is so significant that

medical treatments are not effective, the residual shunt should be eliminated by further embolization, and surgical removal is an alternative option.

37.2.9 Post-procedural Care and Follow-Up

Most patients can be discharged in a few days following the procedure. A pre-discharge imaging study including echocardiography, chest X-ray should be performed to assess the cardiac function and occluder position. Long-term follow-up of treated patients with imaging modality and clinical evaluation is recommended.

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