Basic Concepts and Insights into Aortopulmonary Collateral Arteries in Congenital Heart Diseases

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Introduction

Aortopulmonary collateral arteries (APCAs) are the vascular channels that arise from systemic arteries and supply the pulmonary parenchyma in congenital heart diseases associated with reduced pulmonary blood flow. They are also called systemic to pulmonary collaterals and are embryological remnants of the primitive intersegmental arteries that continue to supply the lungs to compensate for reduced pulmonary arterial blood flow. APCAs commonly arise from the descending thoracic aorta and less frequently from the arch of aorta, branches of subclavian arteries, and abdominal aorta and its branches. Depending upon their size and flow volume, APCAs influence the clinical course, surgical options, and timing of surgical intervention. In this article, we intend to discuss the embryology and studies related to the development and histology of the APCAs, their clinical impact on the underlying condition, and treatment planning. Our focus is on various imaging modalities available in the current era with future prospects, selection of appropriate modality, and endovascular treatment. The surgical options and timing of surgical interventions are also briefly discussed.

Embryological Development, Natural Course, and Associated Conditions

In the early fetal life, primitive intersegmental arteries supply the lung parenchyma prior to the formation of the pulmonary arteries. With the development of the pulmonary arteries, the intersegmental arteries regress and cease to supply the lungs by 50 days postovulation.1 However, they may continue to supply the lungs in case of restricted pulmonary circulation caused by anomalies of right

Abstract

Keywords

► aortopulmonary collaterals
► aortopulmonary collateral arteries
► APCA
► unifocalization
► endovascular APCA embolization

Aortopulmonary collateral arteries are persistent embryological vessels supplying lung parenchyma in various cardiopulmonary diseases with underlying pulmonary hypoperfusion. Their identification and mapping are important because of associated clinical implications and tendency to affect the surgical outcome. This article describes the embryological development and clinical relevance of aortopulmonary collaterals in various congenital cardiopulmonary conditions, along with the significance for treatment planning. Roles, strength, and shortcomings of the various imaging options and image-guided interventions are discussed, with a focus on presurgical planning and preparation, as well as postsurgical management.

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ventricular outflow tract, pulmonary artery, or pulmonary valve. They frequently arise from the descending thoracic aorta at the level of carina; however, they may also arise from the arch of aorta, branches of the subclavian arteries (thoracocervical trunk, internal mammary arteries, lateral thoracic artery, etc.), intercostal arteries, coronary arteries, carotid and vertebral arteries, as well as the abdominal aorta and its branches (Fig. 1–3). On the basis of their origin, they may be classified as branches from bronchial artery (type I), directly from the aorta (type II), or indirectly via aortic major branches (type III). They arborize irregularly and act as peripheral left to right shunts. They may anastomose with the central pulmonary artery in the parahilar location or with the pulmonary artery branches and supply the respective lung segments. When a collateral exclusively supplies a particular part of lung, it is called an "essential (or noncommunicating) collateral" and when the collateral vessel supplies the lung in addition to the respective pulmonary artery branch, it is known as "redundant (nonessential or communicating) collateral." A somewhat contradictory study compares the anatomy of bronchial arteries and APCAs and suggests that APCAs are dilated bronchial arteries rather than noninvolved primitive intersegmental arteries and these may have poor long-term patency in postunifocalization patients. However, this study was limited by unavailability of histological comparison that lacked evaluation of collaterals arising from the coronary arteries. Moreover, a
few authors have criticized this theory by showing favorable long-term patency of the APCAs postunifocalization \[^5,6\] and therefore this theory is not widely accepted.

APCAs can also be classified into nonhypertensive and hypertensive collaterals based on the effect of stenosis.\[^3\] APCAs stenosis occurs in a predictable fashion that can be explained by their underlying histopathology. At their origin, nearly half of the APCAs have a thin elastic wall with larger lumen, while the remainder of APCAs have a more muscular wall at their origin.\[^7\] However, throughout rest of their course, the majority of these APCAs show predominantly muscular walls, with a few showing musculoelastic walls.\[^7\] The APCAs that anastomose with pulmonary artery branches that are not connected to central pulmonary artery show elastic wall with larger lumen, while those that connect to centrally connected pulmonary artery branches show thick muscular wall.\[^7\] APCAs tend to develop stenoses (► Fig. 4) as a natural course, with up to two-thirds of collaterals developing stenoses at least at one site.\[^8\] Common sites of stenosis include near the junction with the aorta or the pulmonary arteries at the hilum.\[^2\] Jia et al\[^9\] and Mainwaring et al\[^10\] found a retroesophageal course of APCAs in approximately two-thirds of patients, and these may demonstrate other characteristics such as—a take-off from the lateral wall of descending thoracic aorta and an origin generally inferior to those of nonretroesophageal APCAs. These retroesophageal APCAs typically arise from the opposite side of aortic arch; the majority (up to 91%) lie posteriorly to the bronchus at the hilum; and nearly half of them show an intraesophageal (coursing through the muscle fibers of the esophagus) course.\[^9,10\] They are more prone to develop stenoses, particularly at the sites where they course posterior or through the esophagus. Histologically, the structure of APA closing is similar to closing of the ductus arteriosus, supporting the fact that both are embryological vessels that tend to involute postpartum, but may persist to support pulmonary circulation.\[^7\] Rarely, they may show aneurysmal dilatation (► Fig. 5) that may rupture and cause pulmonary hemorrhage.\[^11\]

The majority of the patients with APCAs show cardiac morphology of pulmonary stenosis or atresia and ventricular septal defect (VSD). This cardiac morphology leads to reduced pulmonary blood flow and cyanosis that act as triggering factors for noninvolution of the embryological intersegmental arteries. Less frequently APCAs may also be associated with other intracardiac defects that comprise single and double ventricles.\[^12\] Congenital heart diseases with single ventricle morphology, for example, heterotaxy syndrome with unbalanced atrioventricular canal defect with or without total anomalous pulmonary venous return, pulmonary atresia with intact ventricular septum, tricuspid atresia, complex transposition, single left ventricle, and double outlet ventricle, are also associated with APCAs.\[^12\] In one study by Patrick et al, a review of 33 patients with APCAs and single ventricle showed that approximately 50% had heterotaxy syndromes, and it was postulated that a genetic link exists between the two entities. Double ventricle morphology conditions like complete atrio-ventricular canal, corrected transposition of great arteries, double outlet right ventricle, Scimitar syndrome, and complex D-transposition of great vessels are among other congenital heart diseases associated with APCAs.\[^12\]

Fig. 3 Computed tomogram angiography (CTA) shows (A) multiple aortopulmonary collateral arteries (APCAs) from bilateral subclavian artery, APCA from left costocervical trunk (black arrows) courses upwards, turns medially, and descends down and bifurcates to join the right pulmonary artery at hilum. (B) Virtual reconstruction (VRT) image visualized from posterior aspect depicts the same finding. There is narrowing (white arrow) of the proximal segment of the APCA from right costocervical branch. (C) CTA in another patient shows APCA (black arrow) arising from right renal artery and coursing cranially with bifurcation, which ascends toward pulmonary hilum. (D) VRT image in yet another patient shows APCA from middescending thoracic aorta (white arrows) that joins segmental branch of right descending pulmonary artery and another APCA arising from left phrenic artery (black arrow), which ascends cranially with multiple tortuositities and enters mediastinum.

Fig. 4 Computed tomogram angiography in an adult patient with congenital heart disease shows (A) large aortopulmonary collateral artery (APCA) with tight stenosis (white arrow) at its origin from descending thoracic aorta. (B) The anatomy is better defined in virtual reconstructed images in right anterior oblique projection, large APCA with tight ostial stenosis (black arrow).
Clinical Implications of Aortopulmonary Collaterals

Patent APCAs with associated systemic arterial pressures result in pulmonary hyperperfusion by inducing overflow of blood into the pulmonary arteries, while also acting as a peripheral left to right shunt. This leads to pulmonary arterial hypertension and a further pressure overload on an already overburdened right ventricle in case of pulmonary stenosis. This pulmonary hyperperfusion also results in volume overload on the systemic ventricle by increasing pulmonary venous return, leading to congestive cardiac failure. Left ventricular volume overload also leads to progressive aortic annular dilatation resulting in aortic insufficiency and further worsening of left ventricular function. Therefore, high pulmonary blood flow through APCAs should be treated prior to development of pulmonary changes. Increase in pulmonary venous return also has an adverse intraoperative effect as it causes blood flow in the operative field hampering visualization of structures. Artificially created systemic-to-pulmonary shunts are also ligated prior to intracardiac repair for similar surgical impact. However, the presence of good oxygen saturation due to APCAs suggests a good surgical outcome. In contrast, if the blood flow from APCAs is inadequate, this can cause persistent cyanosis and hypoxia. Thus, knowing the significance of APCAs is important for treatment planning in patients with congenital heart defects, as significant APCAs and surgically created systemic-pulmonary shunts require to be occluded prior to definitive surgery. Redundant APCAs in postpalliative surgery patients impede development of the pulmonary artery and branches by continuing to supply systemic blood to the lungs. In patients who are being considered for palliative Glenn shunt, redundant APCAs should be preoperatively embolized to facilitate pulmonary arterial development. In the postoperative period, patent APCAs lead to persistent cyanosis and difficulties with extubation. They may also cause persistent pleural effusion (Fig. 6) and may rupture leading to hemoptysis. In post-Fontan patients who depend on passive venous flow into the pulmonary arteries, APCAs result in increased pulmonary artery pressures and shunting of blood flow resulting in persistent pleural effusions, cardiogenic limb edema and congestive hepatomegaly. APCAs also lead to

Fig. 5 (A) Computed tomogram angiography with virtual reconstructed image shows multiple large aortopulmonary collateral arteries (APCAs) with aneurysmal segment at origin (black arrow). (B and C) Digital subtraction angiogram in another patient shows multiple APCAs from left subclavian artery and one collateral from left internal mammary artery is aneurysmal (white arrow) just before entering the pulmonary circulation.

Fig. 6 Computed tomogram angiography obtained in a child presenting with persistent right-sided pleural effusion after right-sided Glenn shunt shows (A) patent Glenn shunt (black arrows) with right pleural effusion. (B) Multiple aortopulmonary collaterals (arrowheads) were noted to be arising from descending thoracic aorta (DTA) supplying the right pulmonary vasculature. Chest tube noted in situ. White arrows represent the persistent right pleural effusion (C) One large right intercostobronchial trunk (white arrow) noted from DTA. Persistence of flow through aortopulmonary collateral arteries is a known cause of persistent pleural effusion.
volume overload in post-Fontan single (systemic) ventricle patients, leading to increased ventricular end-systolic and end-diastolic volumes, but preserved ejection fraction.\textsuperscript{16}

### Imaging Evaluation

Evaluation of the morphology and functional assessment of APCAs as well as the underlying heart defects serves as the foundation for surgical planning and prognostication. Various noninvasive diagnostic modalities like echocardiography, computed tomography (CT) angiography, and magnetic resonance imaging (MRI) have key roles in modern diagnostic algorithms with each having distinct merits and disadvantages. They are generally complementary to each other and are prescribed depending on the patients’ clinical profile, usually beginning with echocardiography. The challenges pertinent to pediatric population undergoing evaluation include radiation exposure, along with acquisition time and anesthetic risks,\textsuperscript{17} that must be considered while planning an investigation. Chest radiograph remains the basic initial investigation that may provide an important clue to the differential pulmonary vascularity and the associated congenital heart disease.

In the era of noninvasive imaging, CT angiography has emerged as the standard imaging modality for the evaluation of the APCAs. As the patients undergoing CT evaluation for APCA mapping are infants and children, obtaining a diagnostic quality scan, mitigation of motion artefacts, optimization of contrast dose, and maintaining patient comfort with concern for sedation/anesthesia requirement while maintaining an optimal radiation exposure are the primary challenges faced. The CT acquisition and reconstruction techniques have evolved significantly from the time of initial studies where three-dimensional (3D) helical CT demonstrated accurate morphology of pulmonary arteries in complex congenital diseases,\textsuperscript{18,19} to the present day where morphology and functional evaluation are feasible with electrocardiogram (ECG)-gated studies while simultaneously realizing reductions in radiation and intravenous contrast dose along with improvements in acquisition speed. The modern CT scanners provide ultra-high pitch resulting in rapid acquisition, not only alleviating degradation of image quality due to motion artefacts but also reducing the radiation dose to sub-milliSievert level.\textsuperscript{20,21} Faster acquisition also requires contrast nearly the half of that required for routine acquisition.\textsuperscript{22,23} Ultra-high pitch CT angiography (flash mode) uses prospective gating in which systole or diastole is chosen as a target to begin the acquisition. In cases of relatively less complex cardiac diseases, this acquisition usually suffices to provide accurate information of the heart chambers, coronary and major arteries as well as the APCAs. The acquisition may be extended to the level of renal arteries for the diagnosis of APCAs arising from abdominal aorta. Modern iterative reconstruction engines add to the benefits of radiation reduction, providing impeccable contrast resolution. Rarely, patients with complex congenital heart diseases may require evaluation in all phases of cardiac cycle, for which retrospective ECG-gated low-dose CT protocol may be applied. Important information on differential lung perfusion may be obtained using multiphase dual energy CT acquisition. Volume-rendered CT images and 3D printing of the CT data are particularly important in understanding the anatomy, especially for the surgeons, if the surgical ligation or unifoicalization is planned. A noteworthy advantage of CT over MRI is the speed of image acquisition that often eliminates the need for sedation or general anesthesia. A study by Yin et al showed that quantitative analysis of pulmonary arteries and APCAs by CT angiography showed no significant differences to that done by catheter angiography.\textsuperscript{24} 

\begin{table}[h]
\centering
\caption{Important radiology reporting parameters regarding description of APCAs}
\begin{tabular}{|l|}
\hline
Vessel of origin \\
Clock position and vertebral level of APCA origin \\
Description in relation to the carina, bronchus or the esophagus \\
APCA diameter \\
Ostial stenosis \\
Stenosis along the course (length of stenosis and involvement of branching points) \\
Aneurysmal dilatation (with secondary effects including airway compression, if any) \\
Arborization/branching pattern \\
Segmental vascular supply (essential or redundant) \\
Native pulmonary artery anatomy and sizing \\
Lung parenchymal changes \\
\hline
\end{tabular}
\end{table}

Abbreviation: APCAs, aortopulmonary collateral arteries.

MRI is an excellent imaging modality with an advantage of no radiation exposure (\textsuperscript{\textbullet} Fig. 7) as compared to CT angiography.\textsuperscript{25,26} In addition to the use of MR angiography for the evaluation of APCAs, MRI in congenital heart disease can also evaluate ventricular function, pulmonary and aortic flow quantification with calculation of shunt ratio, as well as morphological evaluation. MRI is also advantageous in patients who require repeated evaluation of APCAs, such as in postoperative patients presenting with hemoptysis or persistent pleural effusion or in patients who have persistent symptoms postembolization. Time-resolved MR angiography sequences not only impart dynamic flow details but also determine direction of flow. During the pulmonary arterial phase of time-resolved MR angiography, the pulmonary segments supplied by branches from intrapericardial pulmonary arteries are visualized, while in the systemic arterial phase, the pulmonary segments supplied by the APCAs are seen. This technique can provide excellent images and can delineate up to the fourth-order branches of pulmonary artery.\textsuperscript{27} Noncontrast MR angiography techniques are particularly applicable, whereby contrast administration is precluded due to impaired renal function. A newly devised noncontrast MR angiography sequence, QISS (Quiescent-inflow single-shot, Siemens Aera MRI, Erlangen, Germany), has been studied to image the pulmonary arteries\textsuperscript{28} and...
coronary arteries and may be utilized as a tool to image the APCAs in future. Phase-contrast MRI aids in quantification of flow across a vessel. The blood flow from APCAs contributed to pulmonary flow (Qp) and is considered in calculating total pulmonary blood flow. This was first studied by Grosse-Wortmann et al using phase contrast MRI. They calculated flow across the APCAs directly by evaluating an APCA when possible or in case of complex anatomy of APCAs by subtracting flow within aorta distal to APCA origins from that of the aorta proximal to the origins. They also used total of pulmonary venous return from all pulmonary veins as total pulmonary inflow and systemic flow (Qs) as sum of flow in the aorta distal to the origin of APCAs and flow in the superior vena cava. They also showed that the Qp:Qs ratio thus calculated varied inversely to the intraoperative right ventricular pressure during VSD closure and determined intraoperative decision making and prognosis of VSD closure. Mainwaring et al recently published a study whereby they showed that residual collateral flow in cases of pulmonary atresia and VSD with APCAs correlated to the preoperative oxygen saturation. Whether similar findings can be demonstrated by phase-contrast MRI, it remains a potential area of research. Four-dimensional (4D) flow MRI is a recently devised MRI technique that provides dynamic information of the flowing spins in three dimensions over the entire cardiac cycle. It also provides dynamic flow information through streamlines and velocity vectors alongside more novel parameters, that is, wall shear stress, pulse-wave velocity, turbulence kinetic energy, and pressure difference fields. It is advised for the evaluation of more complex morphology, to determine accurate flow-parameters in any plane and in patients with difficulty in breath-holding.

However, it has not been utilized to evaluate APCAs as yet and this remains a potential research field that may reveal detailed 4D flow patterns, facilitate more accurate quantification of APCAs flow, allow measurement of pressure-gradients across stenotic segments, and enhance calculations of Qp:Qs ratio. Artificial intelligence-based 4D flow quantification is also under development that processes data in short time and may find a utility in APCAs functional evaluation. These modalities are not widely utilized in the current practice due to lack of substantial validating data and cardiac catheterization forms the mainstay for functional evaluation of congenital heart disease and APCAs. For neonates, smaller field of view is required to evaluate smaller caliber vessels and this aspect is yet to be studied and validated. MRI, however, has disadvantages of extended acquisition time and breath-hold that require general anesthesia for acquisition, limiting the widespread utilization of this technique. Although with MRI evaluation of the origin of the coronary arteries is possible, for the evaluation of their course, CT is the preferred noninvasive modality. Artefacts caused by metallic coils also hamper evaluation of the postembolization patients who might require repeat examinations for follow-up or for resurgence of symptoms/complications. The artefacts are particularly marked in previous generation steel coils as compared to modern coils that are made from platinum/alloys. Despite the localized artefacts, the patency of the vessel can be determined with MR angiography.

Cardiac catheterization remains the gold standard for the diagnosis of the anatomy of the collaterals, their arborization pattern, the pulmonary territory they supply, whether or not they communicate with the pulmonary artery branches, presence of stenosis, to look for duct dependent pulmonary blood flow, and for planning endovascular embolization (Figs. 8 and 9). Cardiac catheterization also provides hemodynamic information that is important in assessing the feasibility for single-stage unifocalization and intracardiac repair. Modern angiography equipment provides improved
sensitive and 100% specific branch pulmonary artery less than or equal to 2.5 as 88% patients without APCAs. They used a cutoff value of Z score of significantly small sized branch pulmonary arteries as compared to other angiography. Transposition of great arteries and recommended early catheterization of the APCAs. A study by Wipf et al indicated less sensitivity of echocardiography in postoperative cases of D-skin-dose tracking.

However, the major demerit of this technique is its invasiveness and therefore its utilization is limited only when intervention is planned, oxygen saturation measurement and pressure gradient measurement are indicated.

Echocardiography utilizes the principles of grayscale ultrasound, color Doppler, M-mode, and spectral Doppler to diagnose and characterize congenital heart diseases. A study by Mackie et al showed that echocardiography can be successfully used to diagnose the presence of APCAs. They showed that in patients with APCAs, there were significantly small sized branch pulmonary arteries as compared to patients without APCAs. They used a cutoff value of Z score of branch pulmonary artery less than or equal to 2.5 as 88% sensitive and 100% specific marked of having more than or equal to 1 APCAs. This along with patent ductus arteriosus diameter of less than or equal to 2mm was considered 97% sensitive and 100% specific in their study. However, echocardiography is operator dependent and provides limited evaluation of the coronary arteries, right ventricle, aortic arch/proximal descending thoracic aorta as well as accurate delineation of the APCAs. A study by Wipf et al indicated less sensitivity of echocardiography in postoperative cases of D-transposition of great arteries and recommended early catheter angiography. Selection of an appropriate imaging modality is done on a case-to-case basis keeping in mind the information required and the characteristics of modality (Table 2).

Imaging Mimics of APCAs

APCAs should be differentiated from various imaging mimickers including hypertrophied bronchial artery, patent arterial duct, hemitruncus, acquired collateral arteries, type B malanoscultation, and the venovenous collaterals. Branch pulmonary arteries can be identified by their relationship to the tracheobronchial tree, where they are seen in close adherence to the bronchial tree and following the same course. They can be differentiated from the patent arterial duct that generally arises from the undersurface of the aorta in the region of aortic isthmus, distal to the left subclavian artery origin and generally follows a shorter and nontortuous course, without intervening branches to insert into branch pulmonary artery and may show narrowing at its pulmonary insertion site. Generally, APCAs do not originate from the ascending aorta and can be differentiated from hemitruncus, where one of the pulmonary arteries arises from the ascending aorta. Tracing both ends of the vessel and the attenuation difference on different phases will help in differentiating venovenous collaterals from the APA.

Surgical Management

It is important to ligate significant APCAs at the time of corrective surgery. APCAs are common associations with tetralogy of Fallot morphology conditions and pulmonary atresia with VSDs whereby the modern surgical treatment is single-stage surgery for unifocalization of APCAs and intra-cardiac repair. The single-stage procedure should be performed in early infantile period (as early as 3–6 months) as APCAs tend to develop stenosis as a part of their natural history. Surgery in neonatal period is avoided as it might lead to morbidities due to cardiopulmonary bypass; however, it may be considered in patients with hemodynamic instability due to cyanosis or pulmonary overcirculation or when pulmonary circulation is partially or completely duct dependent.

Unifocalization is surgically performed by incorporating multiple large APCAs and native pulmonary artery if present into a common vascular channel that supplies each lung. The concept of unifocalization of APCAs and incorporating into a single pulmonary artery was first formulated by Haworth and Macartney. Reddy et al first described single stage procedure in early life of patient whereby unifocalization of

Fig. 9 Digital subtraction angiogram in a child with congenital heart disease shows (A) aortopulmonary collateral artery (APCA; black arrow) from right intercostobronchial trunk that travels cranially and after making a hairpin loop, and descends caudally with subsequent joining to pulmonary vasculature. (B) Pulmonary blush (white arrow) is better seen in delayed angiographic phase. (C) In another patient, a large APCA is noted to be arising from right subclavian artery branch, ascends cranially and medially (evident from catheter course) and then descends caudally with filling of right pulmonary artery (white asterisk).
APCAs and intracardiac repair was performed in a single-phase surgery. This showed favorable outcomes and formed a common surgical protocol in modern cardiovascular surgery. One study has shown that surgical unifocalization of the APCAs with intracardiac repair also improves survival and clinical status in patients with single ventricle.

Progressive pulmonary parenchymal damage occurs by hypoperfusion from stenotic APCAs and hyperperfusion from unobstructed APCAs that supply blood at systemic blood pressure. Early single-stage unifocalization with definitive intracardiac repair addresses both these potential causes of pulmonary parenchymal damage. However, Vai- kunth et al have recently demonstrated that patients of tetralogy of Fallot and APCAs can be treated beyond infancy and result in a favorable outcome.

In case of inadequately sized pulmonary arteries, various grafts can be used, for example, cryopreserved pulmonary allograft (Rastelli type correction), polytetrafluoroethylene graft, autologous artery or vein, xenograft pericardium, or autologous pericardium.

Rehabilitation is an alternate and less common surgical approach whereby the diminutive pulmonary arteries are provided with increased circulation by anastomosing them to ascending aorta (Melbourne shunt) or to right ventricle, without addressing the APCAs. A study by Soquet et al showed successful repair in 73% with 10% mortality rate following rehabilitation strategy. Another approach is a combined strategy whereby in the initial stage, pulmonary artery growth is promoted by rehabilitation and subsequently unifocalization is performed prior to corrective surgery.

Surgical ligation of APCAs is a viable treatment option in case of large redundant APCAs that are not occluded despite optimal endovascular embolization. As the APCAs are generally posterior structures, surgical ligation is a relatively complex procedure, especially when the size of collaterals is small. These APCAs are ligated during the cardiac repair and stand-alone surgery for APCAs is not described in view of significant invasiveness. Endovascular embolization is the preferred technique to occlude the APCAs as it is minimally invasive and provides consistent results. It can also be performed in patients who develop long-term complications and provide rapid favorable outcomes as described in the following section.

### Endovascular Treatment
There are several indications for endovascular treatment of APCAs. Because APCAs increase blood flow from lung to left heart during surgery, preoperative treatment of APCAs is important to reduce intracardiac blood flow and provide blood-free surgical field. For augmenting growth of native

### Table 2 Imaging modalities for APCAs evaluation—strengths and limitations

<table>
<thead>
<tr>
<th>Imaging modality</th>
<th>Merits</th>
<th>Demerits</th>
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<tbody>
<tr>
<td>Echocardiography</td>
<td>Diagnosis of underlying congenital cardiac defect</td>
<td>Cannot always directly demonstrate APCAs</td>
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<td></td>
<td>Portability—can be performed on bedside</td>
<td>Operator dependent</td>
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<td></td>
<td>Useful as screening tool</td>
<td>Limited evaluation of right ventricle</td>
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<tr>
<td>CT angiography</td>
<td>Excellent spatial and temporal resolution</td>
<td>Delivers ionizing radiation</td>
</tr>
<tr>
<td></td>
<td>Directly shows accurate anatomy of the APCAs</td>
<td>Susceptible to motion artifacts</td>
</tr>
<tr>
<td></td>
<td>Diagnosis of underlying congenital cardiac defect</td>
<td>Contrast administration risks</td>
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<tr>
<td></td>
<td>Demonstrates status of coronary arteries</td>
<td>May require sedation</td>
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<td></td>
<td>Retrospective ECG-gated CT provides ventricular function information</td>
<td></td>
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<tr>
<td>MR angiography</td>
<td>Provides anatomical details of APCAs</td>
<td>May require sedation or general anesthesia</td>
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<tr>
<td></td>
<td>No risk of radiation exposure</td>
<td>Risk of gadolinium contrast exposure</td>
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<td></td>
<td>Delineation of underlying congenital cardiac defect</td>
<td>Higher susceptibility to motion artefacts</td>
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<td></td>
<td>Gold standard for ventricular function assessment</td>
<td>May not provide spatial resolution to visualized distal vessels</td>
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<tr>
<td></td>
<td>Flow and gradient quantification of APCAs, systemic and pulmonary arteries</td>
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<tr>
<td>Catheter angiography</td>
<td>Gold standard for anatomic information</td>
<td>Invasive modality</td>
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<tr>
<td></td>
<td>Provides dynamic flow information</td>
<td>Risk of radiation exposure</td>
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<td></td>
<td>Possible to measure pressure gradient and oxygen saturation</td>
<td>Risk of contrast administration</td>
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<tr>
<td></td>
<td>Endovascular embolization/flow augmentation possible</td>
<td>Motion artefacts susceptibility</td>
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<tr>
<td></td>
<td></td>
<td>May require sedation or general anesthesia</td>
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</tbody>
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Abbreviations: APCAs, aortopulmonary collateral arteries; CT, computed tomography; ECG, electrocardiogram; MR, magnetic resonance.
persistent cyanosis, persistent pleural effusion (mercury favors VSD closure. pulmonary arterial pressure which if less than 25 mm of operative embolization is also indicated to reduce mean pulmonary pressure.

Table 3: Indications of APCAs embolization in pre- and postoperative settings

<table>
<thead>
<tr>
<th>Preoperative embolization</th>
<th>Postoperative embolization</th>
</tr>
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<tbody>
<tr>
<td>Reduce left heart blood flow</td>
<td>Persistent cyanosis</td>
</tr>
<tr>
<td>Growth augmentation of native pulmonary arteries</td>
<td>Persistent pleural effusion</td>
</tr>
<tr>
<td>Reduce mean pulmonary artery pressure</td>
<td>Hemoptyis</td>
</tr>
<tr>
<td></td>
<td>Difficulty in extubation</td>
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</tbody>
</table>

Abbreviation: APCAs, aortopulmonary collateral arteries.

As not to embolize essential APCAs or large APCAs that can be incorporated into pulmonary artery for viability of the pulmonary segment. Usually 15 out of 20 pulmonary segments' pulmonary arteries should be connected to intra-cardiac pulmonary arteries for successful definitive repair. However, for postoperative cyanosis and hemoptysis, any APCAs suitable for embolization should be occluded to improve patient condition.

For endovascular embolization, various embolization agents are used, for example, Gelfoam pledgets, detachable silicone balloons, metallic coils, and vascular plugs (Figs. 9C, 10 and 11). In modern practice, coils are the most frequently used embolization material. Endovascular coils cause focal arterial thrombosis by platelet aggregation due to entrapment in microfibers and by mechanical occlusion of the vessel. Coil diameter more than one-third to one-half of the artery diameter is selected to avoid migration and more complete occlusion. Endovascular coils available presently are manufactured from platinum, as it is an inert relatively soft metal with low propensity to cause vascular injury. Historically, many earlier coils were made from stainless steel; however, these have been gradually replaced by platinum or alloy coils with microfibers that have higher thrombogenicity and are softer to mold as per the vessel/lesion morphology. Present generation coils are MRI conditional but lead to artefacts in CT and MRI angiography studies rendering postembolization evaluation of the coiled segment difficult. However, this is not the case with catheter angiography and patients who develop symptoms...
postembolization may directly be taken up for catheter angiography depending on clinical condition and severity of symptoms that guide the need for endovascular embolization. Alternate blood supply to the pulmonary segment should be ascertained prior to deployment of coil material. Smaller arteries whereby coil embolization is not possible are embolized using Gelfoam pledgets to cause proximal occlusion. Larger APCAs are better suited for vascular plug embolization to avoid complication of coil migration.\textsuperscript{11} Particulate material embolization is not appropriate in this setting, given the need to preserve the pulmonary parenchyma and avoid any undue damage to the normal lung segment.

Endovascular treatment also includes angioplasty or stenting for stenotic APCAs prior to unifocalization and in postsurgical stenosis. However, the stenotic segments contain large amount of collagen resulting in high resistance to angioplasty and require high balloon pressure as high as 20 atmospheres.\textsuperscript{2} Conventional balloon angioplasty with non-compliant balloon with size not more than 1.5 times of adjacent normal vessel should be selected.\textsuperscript{2} Cutting balloons with metallic blades are used for resistant stenotic lesions and offer smooth dilatation at multiple sites with less chances of elastic recoil. Cutting balloon size of 1.1 times the vessel is selected\textsuperscript{2} and intravascular disruption is usually expected at the site of angioplasty in the check angiogram and is considered important for successful procedure.\textsuperscript{59} An alternate option is endovascular stent placement that has high initial success rates.\textsuperscript{60–62}

**Conclusion**

APCAs play an important role in maintaining viability of the pulmonary parenchyma in congenital cardiopulmonary diseases with impaired pulmonary arterial flow. They arise commonly from descending thoracic aorta and less frequently from other thoracoabdominal arteries. They may undergo stenosis as a part of their natural history and cause pulmonary hypocirculation, while larger APCAs cause pulmonary overcirculation and lead to pulmonary arterial hypertension. It is important to know the origin, number, size, and pulmonary segments supplied by the APCAs so that optimal intervention and surgery can be planned. In modern practice, single-stage surgery in early life is suggested for better surgical outcome. Preoperative and postoperative endovascular interventions play an important role as an adjunct to surgery for further improving the patient’s condition. Various imaging modalities are used for diagnosis and mapping of the APCAs with each modality having specific advantages and limitations. However, technological advances have improved temporal and spatial resolution while also facilitating a decrease in contrast requirements and radiation dose. Also, advancements in 4D flow MRI hold potential to reveal newer insights in functional evaluation of APCAs. Awareness of the embryological development, modern techniques in evaluation, and treatment protocols plays a decisive role in patient management and prognosis.

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**Conflict of Interest**

None declared.

**References**

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