

Percutaneous treatment of anomalous systemic artery to pulmonary venous fistulas in children: Description of three cases and review of the literature

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ABSTRACT

In the normal lung, the only communications between the systemic and pulmonary arterial systems are the connections between the bronchial and pulmonary arteries that occur at the respiratory bronchioles, where pulmonary and bronchial capillaries freely anastomose. Rarely, anomalous connections can occur between normal or aberrant systemic arteries and pulmonary vessels. We performed a comprehensive literature review of all available manuscripts on PubMed and Google Scholar that included a case report or case series with diagnosis of systemic artery to pulmonary venous fistulas who underwent percutaneous treatment. Furthermore, we report three cases of children diagnosed and treated in our Pediatric Cardiology Center.

Keywords: Cardiac catheterization, children, percutaneous treatment, pulmonary fistulas

INTRODUCTION AND BACKGROUND

In the normal lung, the only communications between the systemic and pulmonary arterial systems are the connections between the bronchial and pulmonary arteries that occur at the respiratory bronchioles, where pulmonary and bronchial capillaries freely anastomose.^[1] Rarely, anomalous connections can occur between normal or aberrant systemic arteries and pulmonary vessels. These lesions are acyanotic, and the lack of parenchymal abnormalities together with normal pulmonary arterial and bronchial supply distinguishes systemic arterial supply without sequestration from pseudo and true sequestration [Tables 1 and 2].^[2]

The characteristic features of systemic arterial supply without sequestration (i.e., systemic artery to pulmonary venous fistulas) are resumed in Table 3.^[2]

Although etiology remains unknown, the most likely theory is a persistent embryonic connection between the aorta and the pulmonary parenchyma.^[3] Furthermore, acquired causes exist and include pulmonary suppuration, pleurotomy, and iatrogenic anastomosis.^[4,5] Disease is usually asymptomatic in children, and it may be discovered following the incidental finding of a continuous chest murmur or left ventricular dilation due to the left-to-left shunt, but sometimes, heart failure is the first symptom at the onset.^[6,7] In older children and young adults, cases of massive life-threatening hemoptysis^[8,9] and pulmonary hypertension^[10] have been reported. However, this latter occurs more frequently in case of systemic artery to pulmonary artery (left-to-right) shunts than systemic artery to pulmonary vein (left-to-left) shunts.^[11,12] In addition, endocarditis is considered another possible complication.^[13,14]

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To date, definite indications for treatment still have not been established. However, several authors recommend intervention both in all symptomatic patients and in asymptomatic ones with incidental diagnosis due to the risk of congestive heart failure, endocarditis, and hemorrhagic complications.^[13,15]

Table 1: Definition of types of sequestration complex from Jariwala *et al.*^[2]

Type	Characteristics
True broncho-pulmonary sequestration	Characterized by systemic arterial supply of lung parenchyma with absence of normal bronchial and pulmonary arterial supply Intra lobar - the sequestered lung parenchyma may be included in the substance of lobe Extra lobar - the sequestered lung parenchyma may be anatomically distinct from the remainder of the lung
Pseudo sequestration	The combination of systemic arterial supply to the lung with normal bronchial connections but absent normal pulmonary arterial supply
Systemic to pulmonary vein fistula - non sequestration	It is supplied by the aberrant artery, has no parenchymal or bronchial abnormalities and there is a normal connection with the bronchial tree

Table 2: Classification of arterio-venous fistula/malformation of lungs from Jariwala *et al.*^[2]

Type	Characteristics
Venous - Systemic fistula Fistulous connection between a normal pulmonary arterial branch and the pulmonary venous system.	Causes cyanosis as venous blood is shunted into the systemic circulation
Arterial - Systemic fistula A) Normal systemic artery (e.g., bronchial, internal mammary, intercostal) to pulmonary veins. B) An aberrant systemic artery arising from the descending aorta to pulmonary veins.	Both are acyanotic lesions In type B, pulmonary parenchyma surrounding the fistulous connection may be normal or sequestered

Table 3: Characteristic features of anomalous systemic artery to pulmonary venous fistulas from Jariwala *et al.*^[2]

Lesions can be congenital or acquired, but congenital variety is the most common being diagnosed from newborn to adult age

Hemodynamically significant left-to-left shunts is associated with continuous murmur, bounding peripheral pulses, and left ventricular enlargement

Patients are not cyanotic

Atypical location of the continuous murmur, lack of evidence of increased pulmonary flow and evidence of localized lesions in pulmonary parenchyma, should lead to a suspicion that a left-to-left shunt is present

The bronchial tree, pulmonary artery branching, and affected lung tissue are completely normal

At cardiac catheterization left-to-left shunts are not associated with increased oxygen saturation in the pulmonary artery and may be clearly outlined with use of arteriography

Rapid passage of contrast into pulmonary vein and left atrium at angiography

The same vein also drains the pulmonary circulation

Lesions are often located in the right or left lower lung lobes

Table 4: Review of the literature of anomalous systemic artery to pulmonary venous fistulas percutaneously treated

Patients number	Author	Year of publication	Number of cases	Age and sex	Origin of systemic aberrant vessels	Site of supply in the lungs	Management
1	Brühlmann <i>et al.</i> ^[16]	1998	1	51 years/male	Lower thoracic aorta	Left lower lobe	Embolization with coils
2	Chabbert <i>et al.</i> ^[15]	2002	1	17 years/male	Aorta above the celiac trunk	Basal segments of the right lower lobe	Embolization with coils
3	Kosutic <i>et al.</i> ^[17]	2007	1	3 months/male	Descending thoracic aorta	Right upper lobe	Embolization with coils
4	Singhi <i>et al.</i> ^[8]	2011	1	74 days/female	Descending thoracic aorta	Left lung	Embolization with coils
5	Singhi <i>et al.</i> ^[8]	2011	1	90 days/-	Abdominal aorta	Left lower lobe	Embolization with a vascular plug and coils
6	Jariwala <i>et al.</i> ^[2]	2014	1	7 months/female	Left internal mammary artery and abdominal aorta	Left lung	Embolization with vascular plugs and coils

In the past, surgical ligation of collaterals with or without lobectomy was the therapy of choice. Recently, transcatheter embolization has proved to be a feasible therapeutic alternative.

We performed a comprehensive literature review of all available manuscripts on PubMed and Google Scholar that included a case report or case series with diagnosis of systemic artery to pulmonary venous fistulas who underwent percutaneous treatment. Furthermore, we report three cases of children diagnosed and treated in our Pediatric Cardiology Center of Regina Margherita Hospital in Turin.

CASE SERIES FROM THE LITERATURE

Considering the rarity of this condition, we found only 23 papers of which 5 reporting percutaneous closures, performed in a total of 6 patients [Table 4].

Campbell *et al.*,^[18] Scott and Perry (1969),^[5] and Ernst and Bruschke^[19] reported systemic artery to pulmonary vein fistulas with left-to-left shunt first. Later on, other isolated case reports, mostly congenital, were published.

In the past, surgery has been the treatment of choice, but in 1998, Brühlmann *et al.* described the first therapeutic transarterial embolization with coils of a systemic artery to pulmonary vein fistula in a 51-year-old man presenting with massive hemoptysis. In their case report, a large anomalous artery arising from the lower thoracic aorta and supplying the basal lung segments of the left lower lobe was diagnosed with a computed tomography (CT) scan.^[16]

Since then, other authors reported successful percutaneous embolization of this kind of lesions.

Jariwala *et al.* reported the case of a 7-month-old girl who was referred to the hospital by a general practitioner with failure to thrive and suspected congenital heart disease.

CT angiography showed anomalous systemic feeders of the left lung from the branches of the left mammary and left axillary arteries and from an aberrant vessel originating from the upper abdominal aorta. Collaterals were embolized using vascular plugs and coils.^[2]

Chabbert *et al.* described the case of a 17-year-old boy who was admitted to the hospital for chest pain. CT scan confirmed the presence of an aberrant aneurysmal artery arising from the aorta above the celiac trunk and supplying the basal segments of the right lower lobe without any parenchymal abnormality. Aberrant artery was successfully embolized using several coils.^[15]

Kosutic *et al.* reported the case of a 3-month-old infant with signs and symptoms of vascular tracheal compression and congestive heart failure. Aortography showed two major aberrant arteries arising from the

descending thoracic aorta and entering the right upper pulmonary lobe. One of them was successfully embolized with two coils and the other one spontaneously closed after cardiac catheterization.^[17]

Singhi *et al.* reported the case of a 74-day-old term baby who had tachypnea, feeding difficulty, and failure to thrive. An anomalous artery from the descending aorta to the lower segment of the left lung was diagnosed and then embolized with three coils.

They also reported the case of a 90-day-old term baby presenting with tachypnea and feeding difficulty. Two collaterals arising from the abdominal aorta and supplying the lower lobe of the left lung were identified and then embolized using a vascular plug and five coils.^[8]

CASE SERIES FROM OUR CENTER

Case #1 is a 5-year-old healthy boy, who was hospitalized at the age of 3 years for an episode of right basilar bronchopneumonia, treated with antibiotic therapy. During hospitalization, a continuous murmur was detected at the right hemithorax. Echocardiography was normal; therefore, he underwent CT scan that showed an aberrant artery connecting the mean thoracic aorta to the laterobasal segment of the right lung, without any parenchymal abnormality. Because of the potential hemorrhagic risk and development of pulmonary hypertension, we decided to embolize the vessel. Hence, a conventional arterial catheterization (femoral, 5F) was performed. Aortography showed a large collateral arising from the thoracic aorta, nourishing the inferior lobe of the right lung, and an additional minor collateral arising from the right mammary artery [Video 1].

Selective angiography of the aberrant artery showed a large left-to-left shunt that was successfully embolized with a vascular plug (Amplatzer vascular Plug1 8 mm).

Selective angiography of the right mammary artery confirmed perfusion of the right inferior pulmonary lobe by an aberrant vessel that was occluded with a second vascular plug (Amplatzer vascular Plug1 6 mm).

Postembolization aortography showed complete occlusion of the aberrant artery from the aorta and scarce residual flow through the mammary artery beyond the plug [Video 2].

Control cardiac catheterization performed 7 months later showed the development of other additional collateral vessels arising from the thoracic aorta and the right mammary artery nourishing the inferior lobe of the right lung [Video 3].

All anomalous vessels were embolized with vascular plugs of various sizes (Amplatzer vascular Plug1 8 and 10 mm large and Plug4 5 and 6 mm large). Postembolization

arteriographies showed scarce residual flow in the feeding arteries beyond the plugs [Video 4].

At the last clinical follow-up, 8 months later, the child was in good health and he did not assume any drug.

Patient #2 is an ex very low birth weight preterm male (birth weight 550 g) with a history of a severe bronchopulmonary dysplasia and respiratory failure.

After hospital discharge, he was readmitted at the age of 6 months for respiratory failure. Cardiac examination showed pulmonary hypertension, mitral and tricuspid valve insufficiency, and left ventricular enlargement. He underwent thoracic CT scan that showed partial lung consolidation with widespread images of ground glass.

At the age of 3 years, clinicians noticed a murmur at the right hemithorax and therefore he underwent a diagnostic 6F femoral artery approach catheterization. Aortography showed several collateral vessels from the thoracic aorta and the right mammary artery to the entire right lung [Video 5].

Occlusion of three collaterals arising from the aorta was performed using vascular plugs (Amplatzer vascular Plug4 7 mm and Amplatzer vascular Plug1 10 mm) and microparticles, while the hypertrophic mammary artery was embolized with eight coils and a vascular plug (Amplatzer vascular Plug1 10 mm).

Final aortography showed poor residual flow to the right lung [Video 6].

Control cardiac catheterization performed 10 months later still showed large anomalous vascularization to the right lung arising from the thoracic aorta and right subclavian artery [Video 7].

We embolized two collaterals from the subclavian artery: one with a plug (Amplatzer vascular Plug4 7 mm) and one with a coil and two collaterals from the thoracic aorta: one with a plug (Amplatzer vascular Plug4 6 mm) and one with a coil.

Although final aortography showed moderate residual flow to the right lung, we decided not to prolong the length of the examination, considering that the child was very fragile.

At the last clinical follow-up, 8 months later, parents reported overall good health with improved respiratory condition. At physical examination, murmur was still hearable and left ventricle appeared still dilated at echocardiography. He will undergo a new planned cardiac catheterization in the next months.

Patient #3 is a 3-year-old boy who entered into the emergency room with hemoptysis. Thoracic CT scan showed lung parenchymal consolidation of the anterolateral segment of the left lower lobe with images of vascular malformations in its structure.

Therefore, he underwent a 5F femoral arterial catheterization which showed two large collateral vessels from the thoracic aorta to both lungs. The left one gave rise to a complex arteriovenous malformation with a large connection with the inferior left pulmonary vein [Video 8].

We performed embolization of both collaterals with two plugs (Amplatzer vascular Plugs4 5 and 6 mm).

Final aortography showed no residual vascularization through the collaterals to the lungs [Video 9].

The patient was discharged in good clinical conditions.

All procedures we described were uneventful.

DISCUSSION AND CONCLUSIONS

Anomalous systemic arterial supply to normal lungs represents a subtype of a rare group of bronchovascular anomalies which can have varied presentation. Although predominately reported in the adult population, pediatric case reports are also available in the literature.

In the past, surgical ligation of collaterals with or without lobectomy was the therapy of choice. Recently, transcatheter embolization has proved to be a feasible therapeutic alternative in these cases without sequestration.

In our opinion, the best approach in case of suspicion is to perform contrast CT first in order to confirm the diagnosis and plan the best therapeutic strategy, also considering the low X-rays and contrast doses needed with the newest CT scanners.

Then, the patient can undergo cardiac catheterization aiming at the closure of as many collaterals as possible.

As reported, the Amplatzer vascular plugs are largely used for transcatheter occlusion of abnormal vessel communications with a high success rate. The only disadvantage may be the need for relatively large catheters (5F at least) in small children.^[20] Controlled-release coils are other common devices consisting of a metallic coil with attached fibers that increase their occlusive effect. Their advantage is that they can be implanted through 4F catheters.^[21,22]

In our cases, we successfully used plugs for larger fistulas and coils for smaller ones.

In the literature, immediate and short-term follow-up results proved good, but long-term postembolization data are scarce. Based on our experience, some embolized anomalous connections can show persistent residual flow, as well as new collaterals can develop over time, suggesting that these lesions likely represent a heart-lung disease more than an isolated anomaly of one or more vessels.

For these reasons, a careful follow-up remains mandatory, and in case of disease relapse, a new interventional cardiac catheterization is needed, in order to prevent the severe complications reported in untreated patients.

We recommend a control cardiac catheterization after 6–12 months, preceded by a new contrast CT examination in case of doubts about the anatomy or extensive disease.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient's guardian has given consent for patient's images and other clinical information to be reported in the journal. The patient's guardian understands that patient's name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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