

# Therapeutic Embolization of Anomalous Systemic Arterial Supply to the Normal Basal Segments of the Left Lower Lobe — A Safe Alternative to Surgery

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Anomalous systemic arterial supply to the normal basal segments of the left lower lobe is a rare congenital anomaly. Although most cases are asymptomatic, treatment is recommended for all patients with or without symptoms because of their potential risk for hemoptysis due to pulmonary hypertension and heart failure due to systemic artery-to-pulmonary vein shunt. There are several surgical procedures available for this rare disease, including lobectomy, ligation or anastomosis of the anomalous systemic artery. Transarterial embolization is a new and semi-invasive technique compared with surgical treatment. Here we describe one 3 month-old female infant with congestive heart failure and one 13 year-old girl with mitral regurgitation and hemoptysis. Computed tomography and angiography demonstrated anomalous systemic artery supply to the left lower lung without coexistence of pulmonary sequestration in both cases. Both patients underwent successful coil embolization of the anomalous artery. Our experience suggests that transarterial embolization could be a safe and effective alternative method to surgery.

**Key Words:** Therapeutic embolization • Pulmonary sequestration • Anomalous systemic arterial supply

## INTRODUCTION

Anomalous systemic arterial supply to the normal basal segments of the left lower lobe (systemic arterialization of the lower lobe without sequestration) is a rare congenital form of bronchopulmonary-vascular anomaly. Such an anomalous systemic arterial supply has been reported as an isolated lesion because the involved lung has normal bronchial connections and pulmonary arteries, which distinguishes this entity from classic pulmonary sequestration.<sup>1,2</sup> Angiography is the most useful method to establish the exact diagnosis and accurately delineate

the vascular anatomy. There are several surgical procedures available for this rare disease. In most reported cases, the anomalous vessel was ligated and the involved lung parenchyma was resected.<sup>3</sup> Other operative procedures, including anastomosis between the anomalous artery and pulmonary artery or simple ligation of the anomalous artery, have been reported.<sup>4</sup> Transarterial embolization is a new and semi-invasive technique compared with surgical treatment. This report describes our experience of two cases treated successfully with coil embolization of the anomalous systemic artery. This experience suggests that transarterial embolization could be a safe and effective alternative method to surgery.

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## CASE REPORT

### Case 1

This 3-month-old female infant was born at the gestational age of 38 weeks by normal spontaneous delivery without complicated pregnancy. She was referred to our

hospital for cardiac evaluation due to respiratory distress and a heart murmur. Growth and development were normal. On physical examination, a systolic ejection murmur, grade III/VI, was audible loudest at the left lower parasternal area. The liver was 3 cm below the right costal margin, and mild bilateral subcostal retraction was found. Two-dimensional echocardiography revealed dilation and hypertrophy of the left ventricle and left atrium. The intracardiac anatomy otherwise appeared normal. Color Doppler echocardiography demonstrated mild mitral regurgitation. In addition, an increased velocity of flow from the pulmonary vein to the left atrium and a retrograde flow during diastolic phase over the descending aorta were also noted. An abnormal vessel was also seen from the mid descending aorta to the left lower lung. These findings suggested an abnormal vascular communication between the descending aorta and the pulmonary circulation. The chest computed tomography (CT) showed neither associated mass nor structural abnormality of the pulmonary parenchyma and bronchus. Compared with the opposite side, however, a hypervascularity in the basal segments of the left lower lobe was noted (Figure 1). The magnetic resonance angiography (MRA) showed a delayed perfusion of the left lower lobe synchronized with the descending aorta and an abnormal vessel arising from the descending thoracic aorta (Figure 2). The hemodynamic data of the cardiac catheterization are shown in Table 1. Aortography and selective arteriography revealed an anomalous tortuous artery arising from the distal thoracic aorta entering the basal



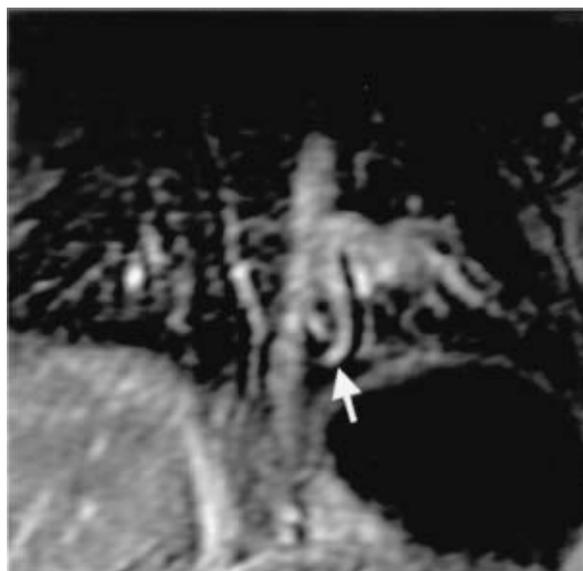
**Figure 1.** Chest computed tomography showed the “Medusa’s hair”-like crowded and dilated anomalous arterial branches penetrating into the basilar segments of the normal left lower lobe.

segments of the left lower lobe (Figure 3A). There was a normal pulmonary capillary phase, and the venous drainage was into the left atrium by the inferior pulmonary vein, thereby excluding a direct fistula between the aberrant systemic artery and the pulmonary veins. Pulmonary angiography showed a normal left pulmonary artery with an absent artery supply to the basal segments. The aberrant artery was then embolized using five coils (5 mm in diameter; Cook Incorporated; Bloomington, Indiana) without complication. Selective angiography after embolization indicated almost complete occlusion (Figure 3B). She had an excellent recovery from the therapeutic embolization and has remained free from heart failure since then. During follow-up, the size of the left atrial and ventricular chamber returned to normal, with-

**Table 1.** Hemodynamic data of bilateral catheterization

Data	Case 1	Case 2
RVSP (mmHg)	30	27
PASP (mmHg)	38	25
LVSP (mmHg)	94	115
AoSP (mmHg)	87	112
Qp/Qs	1.8	1.2

RVSP = right ventricle systolic pressure; PASP = pulmonary artery systolic pressure; LVSP = left ventricle systolic pressure; AoSP = aortic systolic pressure.



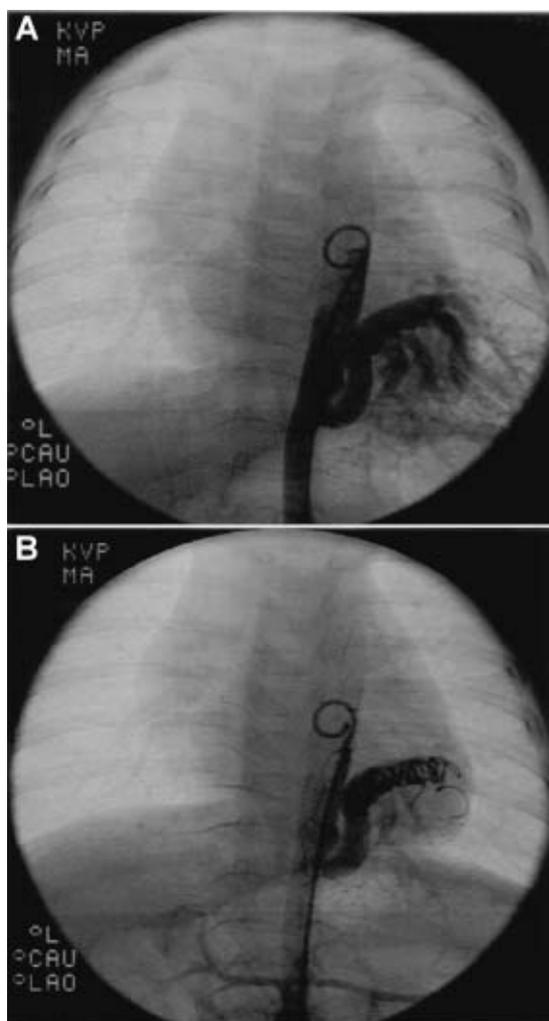
**Figure 2.** The coronal magnetic resonance angiography demonstrated an anomalous vessel arising from the descending thoracic aorta (arrow).

out mitral regurgitation on echocardiography. The technetium-99 m lung perfusion scan revealed no blood flow over the basal segments of the left lower lobe. At follow-up 6 months later, she was at normal growth and development without complication.

## Case 2

A 13-year-old girl was admitted to our hospital with the first episode of intermittent hemoptysis for one week. She had been diagnosed as congenital mitral regurgitation since 4 months old and received regular follow-up at our outpatient department without other clinical symptoms. The physical examination revealed a systolic ejec-

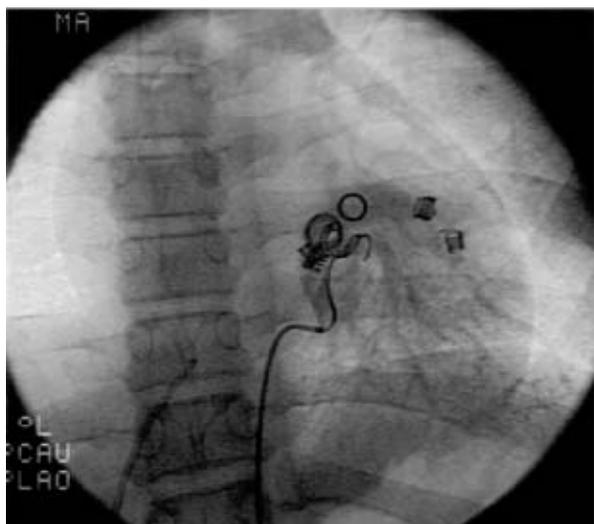
tion murmur, grade III/VI, at the left lower parasternal border. The chest roentgenogram showed increased infiltrating density and vascular shadows over the left lower lung field. Two-dimensional and color Doppler echocardiography revealed dilation of the left ventricle and atrium combined with mitral regurgitation. The chest high-resolution CT showed a hypervascularity in the basal segments of the left lower lobe without structural abnormality of the parenchyma and bronchus. The MRA showed an abnormal vessel from the descending thoracic aorta. Integrated color Doppler echocardiography showed similar findings. The hemodynamic data of the cardiac catheterization are shown in Table 1. The thoracic aortography and selective angiography revealed an anomalous artery arising from the distal thoracic aorta to the left lower lobe, and the venous drainage returned normally to the left atrium by the inferior pulmonary vein. Pulmonary angiography demonstrated a normal left pulmonary artery with an absent artery supply to the basal segments. We used six coils (8 mm diameter) to embolize the supplying artery without complication (Figure 4). Hemoptysis stopped completely after therapeutic embolization. During follow-up, the echocardiography showed improved mitral regurgitation and decreased size of left atrial and ventricular chamber. We performed repeated catheterization 6 months later. When residual arterial supply was found, re-embolization was done smoothly with two coils (8 mm diameter). During the follow-up period of 18 months, she has been well without any complications.



**Figure 3.** (A) Thoracic aortogram demonstrated an anomalous artery arising from the distal thoracic aorta entering the basal segments of the left lower lobe. (B) The aberrant artery was embolized by five coils and indicated almost complete occlusion after embolization.

## DISCUSSION

Anomalous systemic arterial supply to normal segments of the lung is a rare congenital anomaly. Although the etiology of systemic arterial supply to normal lung is still unknown, it is generally believed that this might result from an abnormal persistence of the embryonic connection between aorta and pulmonary parenchyma.<sup>5</sup> This disease was previously classified as Pryce Type I sequestration.<sup>6</sup> However, the absence of parenchymal abnormalities and a normal bronchial supply clearly distinguish this systemic arterial supply from true pulmonary sequestration, characterized by a mass of nonfunctioning lung tissue separated from the normal bronchopulmonary tree and vascularized by an aberrant systemic artery.<sup>3</sup> In



**Figure 4.** Thoracic aortography revealed an anomalous artery arising from the distal thoracic aorta to the left lower lobe. The anomalous artery was embolized with six coils without complications.

addition, it is also different from pulmonary sequestration because no accompanying anomalies have been described in this systemic arterialization.<sup>7</sup> As in both of our cases, the chest CT demonstrated normal bronchial trees and lung parenchyma without coexistence of pulmonary sequestration, and showed the crowded and dilated anomalous arterial branches. The MRA and angiography also confirmed an anomalous artery arising from the distal thoracic aorta entering the basal segments of the left lower lobe. Therefore, if this anomaly is kept in mind, the diagnosis is not so difficult to make because of the characteristic image findings. Yamanaka et al, in reviewing 16 cases of systemic arterial supply without sequestration, found that male gender, left lower lobe, aberrant artery arising from descending thoracic aorta, absence of pulmonary blood flow to the basal segments, and normal pulmonary venous drainage were predominant clinical features.<sup>4</sup> Both our cases were also compatible with the general trend, except that both were females.

Symptoms of systemic arterialization of the left lower lobe without sequestration are variable. Most patients, especially children, are often asymptomatic in the systemic arterialization to lung. The anomaly may be discovered following the incidental finding of a heart murmur over the left anterior chest or over the back.<sup>8</sup> Occasionally, the shunt is sufficiently large to produce left-sided cardiac overload and congestive heart failure, as in our first case.<sup>2</sup> Therefore, if a continuous murmur

over the precordium and the lower posterior thorax or unilateral increase in pulmonary blood flow is noted, this diagnosis should be suspected.<sup>4</sup> Massive hemoptysis seems to be an unusual manifestation in the reviewing previously reported cases.<sup>3</sup> Pathological examination of these anomalous arteries documented that the walls had elastic laminae within their medium but not muscular, unlike bronchial arteries. Such a wall could not stand the high pressure of systemic artery.<sup>9</sup> This might explain the etiology of hemoptysis in the anomalous systemic arterial supply to the normal lung, just as in our second case.

Treatment is recommended for all patients with this anomaly with or without symptoms because of its potential risk for hemoptysis due to pulmonary hypertension and heart failure due to systemic artery to pulmonary vein shunt.<sup>2</sup> For correction of the shunting, surgical intervention is performed in most reported cases, including simple ligation of aberrant systemic artery, anastomosis of the pulmonary artery or simple lobectomy.<sup>1</sup> For pulmonary sequestration, most authors agree to treat the sequestered lung parenchyma because of its propensity for recurrent infection.<sup>7</sup> However, in contrast to pulmonary sequestration, if the parenchyma is normal, as one might encounter in systemic arterialization to lung, only a ligation of the systemic vessel by surgical ligation or coil embolization may be sufficient.<sup>3</sup>

Transarterial embolization has recently been well established in the management of pulmonary sequestration.<sup>10</sup> In reviewing the English-language literature to date, six cases of therapeutic embolization by coils to a systemic arterialization of lung without sequestration in pediatric group have been described (Table 2). Because of less complication and shortened recovery time, the embolotherapy is considered as a semi-invasive and effective treatment compared with surgical intervention, at least in patients who have a dual (systemic and pulmonary artery) supply to the affected segment.<sup>3</sup> In most previously reported cases, if the anomalous artery represented the sole source of blood flow to the involved pulmonary parenchyma, simple lobectomy or segmentectomy was done. We first tried to perform the transarterial embolization in our two patients with an absent pulmonary arterial supply to the affected segment. No postembolization complication was demonstrated by clinical and angiographic follow-up. Also, the clinical improvement in our both patients after the procedure is

**Table 2.** Review of the English literature published to date of therapeutic embolization of anomalous systemic arterial supply to normal lower lobe of the lung

Reference/ Year	Age (years)	Sex	Clinical manifestations	Anomalous artery origin/supply	FU/Complication
1. Chabbert <sup>11</sup> /2002	17	M	Chest pain	Abd A/ RLL	12 months/N
2. Abdulhamid <sup>12</sup> /2004	7	F	Hemoptysis	DTA/RLL	6 years/N
	13	M	Chest pain/Hemoptysis	LIMA & RICA/Right lung	18 months/N
	15	F	Hemoptysis	DTA/RML & RLL	18 months/N
	7	M	Hemoptysis	Aorta/Right lung	2 years/N
	10	M	Hemoptysis	Aorta/ not mention	9 months/N
3. Present/2005	3	F	CHF	DTA/LLL	6 months/N
	13	F	Hemoptysis	DTA/LLL	18 months/N

Abd A = abdominal aorta; CHF = congestive heart failure; DTA = descending thoracic aorta; LLL = left lower lobe; LIMA = left internal mammary artery; RICA = right intercostals artery; RLL = right lower lobe; RML = right middle lobe; PA = pulmonary artery.

clear evidence of the reversal of the adverse effect of abnormal arterial flow on their cardiac and vascular status.

Including our report, eight cases of therapeutic embolization to a systemic arterialization of normal lung in pediatric group have been described in the English literature (Table 2). To the best of our knowledge, we are the first to use therapeutic embolization in cases of anomalous systemic arterial supply to the normal basal segments of the left lower lobe with an absent pulmonary arterial supply to the basal segments.

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## 用心導管栓塞術治療支配左下肺葉的不正常主動脈分支： 一種安全且非外科手術的治療方法

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支配正常左下肺葉的不正常主動脈分支是一種少見的先天性異常。雖然大多數的病人並沒有出現症狀，但建議不管是否出現症狀皆需治療，因為有併發肺高壓咳血及心衰竭的潛在危險。目前已有數種外科治療的方式來處理這種少見的疾病，包括肺葉切除、結紮或吻合這些不正常血管。用心導管栓塞不正常主動脈分支是一種侵襲性較小且非外科手術的治療方法。我們陳述兩個案例，分別是呈現心衰竭的三個月女嬰和二尖瓣回流併有咳血的十三歲女孩。兩例的電腦斷層和血管攝影皆顯示有支配正常左下肺葉的不正常主動脈分支，並被心導管栓塞術治療成功。我們建議心導管栓塞術是一種安全且和外科處理一樣有效的替代方法。

**關鍵詞：**心導管栓塞術、游離肺、不正常主動脈分支。