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THE CARDIOPULMONARY  
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*Chest* 2002;122:1471-1477

DOI: 10.1378/chest.122.4.1471

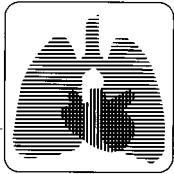
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A M E R I C A N C O L L E G E O F  
 C H E S T  
P H Y S I C I A N S



# Isolated Unilateral Absence of a Pulmonary Artery\*

## A Case Report and Review of the Literature

A. Derk Jan Ten Harkel, MD, PhD; Nico A. Blom, MD; Jaap Ottenkamp, MD, PhD

**Objective:** The purpose of the present study was to determine the symptomatology, diagnostic procedures, and therapeutic strategies of patients with an isolated unilateral absence of a pulmonary artery (UAPA).

**Background:** Isolated UAPA is a rare anomaly. Some case reports exist, but the best diagnostic and therapeutic approaches to these patients remain unclear.

**Methods:** A retrospective analysis was made of 108 cases reported between 1978 and 2000. The database of the National Library of Medicine (MEDLINE) was used to identify cases that were published in any language from 1978 onward.

**Results:** Of the 108 patients identified, 14 were asymptomatic. The median age was 14 years (range, 0.1 to 58 years). Most patients had symptoms such as frequent pulmonary infections (37%), dyspnea or limited exercise tolerance (40%), or hemoptysis (20%). Pulmonary hypertension was present in 44% of the patients. Surgical procedures were performed in 17% of patients, and the overall mortality rate was 7%.

**Conclusion:** Only a few patients with isolated UAPA remain asymptomatic during follow-up. The diagnosis can be made by chest radiograph, echocardiography, CT scan, and MRI. Hilar arteries can be shown by cardiac catheterization and pulmonary venous wedge angiography. This is important since revascularization may improve pulmonary hypertension. The avoidance of high altitudes and pregnancy may further improve outcomes.

(*CHEST* 2002; 122:1471-1477)

**Key words:** diagnosis; pulmonary artery agenesis; pulmonary hypertension; symptoms; therapeutics; unilateral absence of pulmonary artery

**Abbreviations:** HAPE = high-altitude pulmonary edema; UAPA = unilateral absence of a pulmonary artery

Congenital unilateral absence of a pulmonary artery (UAPA) is a rare anomaly, most frequently accompanied by cardiovascular anomalies such as tetralogy of Fallot<sup>1-9</sup> or septal defects.<sup>2,10-12</sup> Other congenital heart defects that have been associated with UAPA are the following: coarctation of the aorta, either isolated<sup>13</sup> or in combination with a ventricular septal defect<sup>14</sup>; subvalvular aortic stenosis<sup>1,15</sup>; transposition of the great arteries,<sup>16</sup> either isolated or in combination with ventricular septal defect<sup>1</sup> or pulmonary stenosis<sup>10</sup>; Taussig-Bing malformation and coarctation<sup>17</sup>; congenitally corrected transposition and pulmonary stenosis<sup>18</sup>; and scimitar syndrome.<sup>19-22</sup> Although less frequent, UAPA also may occur as an isolated finding. Although some patients with isolated UAPA are completely asymptomatic,<sup>23</sup> others may have severe pulmonary hypertension and congestive heart failure.<sup>24,25</sup>

After the first case report in 1868,<sup>26</sup> many case reports and a few reviews have been published,<sup>27,28</sup> but we have found no review of isolated UAPA published after 1978. Since that time, however, major technological advances have been made, and the role of echocardiography, especially in the diagnostic workup of patients with congenital heart defects, has dominated. Because many congenital heart defects associated with UAPA can have the same signs and symptoms as isolated UAPA (eg, recurrent respiratory infections and congestive heart failure), we decided to study patients with isolated UAPA only.

In the present study, we reviewed all cases in which isolated UAPA had been reported from 1978 onward, as well as including a recent case report from our own institute. We hypothesize that, at present, the diagnosis of isolated UAPA is usually made before the onset of pulmonary hypertension or severe symptoms of hemoptysis. We also propose a diagnostic workup for patients with suspected UAPA.

### MATERIALS AND METHODS

All patients reported in the literature since 1978 with isolated UAPA (left or right pulmonary artery) were included. More specifically, patients with associated congenital cardiac anomalies

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Manuscript received September 5, 2001; revision accepted May 16, 2002.

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were excluded, except those with minor (clinically insignificant) left-to-right shunt without pulmonary hypertension.

The database of the National Library of Medicine (MEDLINE) was used to identify cases that had been published in any language from 1978 onward. The key words "pulmonary artery agenesis" or "absent pulmonary artery" were used. The reference lists of all identified journal articles also were reviewed.

From the case reports, we extracted the diagnostic procedures, the symptomatology, and the therapeutic strategies applied. The most frequently performed diagnostic procedures were the following: ventilation-perfusion scanning; and cardiac catheterization, including pulmonary venous wedge angiography, echocardiography, and CT scanning or MRI. We also extracted more details about the presence or absence of recurrent pulmonary infections, dyspnea or decreased exercise capacitance, hemoptysis, and pulmonary hypertension. Since not all reports gave sufficient detail about the symptoms or procedures performed, the percentages reported here refer only to those cases with sufficient details.

### CASE REPORT

A 3-year-old girl was apparently completely healthy on hospital admission. There were no signs of recurrent respiratory infections, no dyspnea, no hemoptysis, and a normal exercise tolerance. A chest radiograph, which had been performed for routine tuberculosis screening, showed a decreased pulmonary vasculature of the left lung and a displacement of the heart to the left. A soft continuous murmur was heard at

the upper left sternal border. Otherwise, the findings of the physical examination were normal. Echocardiography showed a right descending aortic arch, small persistent ductus arteriosus, and the absence of the left pulmonary artery. No left pulmonary venous return was detected by color Doppler echocardiography. A perfusion scintigram showed the absence of perfusion in the left lung. Cardiac catheterization confirmed the diagnosis of an absent left pulmonary artery, a small hemodynamic insignificant ductus from the right pulmonary artery, and a right descending aortic arch (Fig 1). There was no pulmonary hypertension. On venous wedge angiography, no left pulmonary arteries were found. Since there were no symptoms, we decided to carefully follow this child and not perform any interventions at that time.

### RESULTS

Including our own case, we found a total of 108 patients with UAPA of the right or left pulmonary artery without associated cardiac anomalies. These cases were described in 62 different articles.<sup>1,2,10,16-18,20,23-25,29-30</sup> The median age of these patients was 14 years (range, 0.1 to 58 years). Thirteen patients (12%) were infants < 1 year of age.

In 37% of patients (40 of 107 patients), the left pulmonary artery was absent, while the right was absent in 63% of patients (67 of 107 patients). One report<sup>29</sup> was



FIGURE 1. Pulmonary angiogram in the anterior view showing the absence of the left pulmonary artery.

inconclusive about the involved side. In only three cases, the aortic arch was on the same side as the absent pulmonary artery.<sup>16,30</sup>

Symptoms were chest pain,<sup>35,36</sup> pleural effusion,<sup>35</sup> and recurrent pulmonary infections in 37% of patients (35 of 94 patients), dyspnea or exercise limitations in 40% of patients (36 of 92 patients), and hemoptysis in 20% of patients (18 of 92 patients). Pulmonary hypertension was found in 44% of patients (25 of 57 patients), while high-altitude pulmonary edema (HAPE) was present in 12% of patients (7 of 57 patients) [Table 1].

In 14 asymptomatic patients, the diagnosis of UAPA was made after abnormal results from a chest radiograph, which had been made for the following reasons: a tuberculosis survey<sup>23,48</sup>; a routine chest radiograph for induction into the army<sup>16</sup>; routine examination before an operation<sup>17</sup>; or for scoliosis.<sup>32</sup> Six patients received diagnoses after experiencing HAPE,<sup>45–47</sup> and in three patients the diagnosis was made during pregnancy<sup>37</sup> or immediately after pregnancy<sup>38,39</sup> because of respiratory difficulties.

In all patients, different investigations were made, including CT scans,<sup>31–33</sup> MRI scans,<sup>32–34</sup> and bronchography.<sup>16,23</sup> A perfusion scan of the lungs was performed in 59% of patients (55 of 94 patients), echocardiography in 34% of patients (34 of 100 patients), and cardiac catheterization in 90% of patients (98 of 108 patients) [Table 1]. In four patients, a pulmonary venous wedge angiogram was made to identify hidden pulmonary arteries. The time period had a marked influence on the usage of echocardiography. From 1990 onward, echocardiography was used in 65% of cases compared with only 15% of cases between 1978 and 1990. The same trend was seen for the use of CT scanning or MRI. From 1990 onward, CT scanning or MRI were used in 52% of cases compared with only 6% of cases between 1978 and 1990.

Therapeutic approaches also differed. In nine cases (8% of patients), a pneumonectomy or lobectomy was performed for reasons of recurrent hemoptysis or intractable

pulmonary infections (Table 2). The revascularization of hidden pulmonary arteries was performed in eight patients (7%) [Table 2].

One of the three patients with pulmonary hypertension during pregnancy died after delivery.<sup>38</sup> Overall, seven patients (7%) died of the following causes: massive pulmonary hemorrhage<sup>40,41</sup>; right heart failure<sup>42,43</sup>; respiratory failure<sup>44</sup>; pulmonary hypertension<sup>38</sup>; or HAPE.<sup>45</sup>

## DISCUSSION

The first report on UAPA was published in 1868.<sup>28</sup> The first review of 98 cases, of which 32 were isolated UAPA, was published in 1962.<sup>28</sup>

Since many patients with UAPA can remain asymptomatic for a long period, the actual prevalence of UAPA is difficult to establish. Some reports state that 30% of patients are asymptomatic.<sup>16,49</sup> In the present review, only 14 patients were found to be asymptomatic, while another 9 patients were unmasked by predisposing factors for pulmonary hypertension such as pregnancy or high altitude.<sup>37–39,45–47</sup> In studying the case records, there may, however, be some bias toward the most symptomatic patients. One of the reasons for UAPA remaining unrecognized for a considerable time is that symptoms are not always specific, leading to a considerable delay between the onset of symptoms and the final diagnosis.<sup>28</sup> In the present study, we found patients with a time delay of > 30 years. For example, one patient, a 58-year-old woman, had experienced episodes of fever, hemoptysis, and pulmonary infiltrates for almost 30 years before the diagnosis was made.<sup>35</sup> In another patient, multiple protracted pulmonary infections since childhood had preceded the final diagnosis at the age of 39 years.<sup>50</sup>

The best estimation of prevalence came from a study by Bouros and colleagues<sup>16</sup> that included 600,000 participants in whom chest radiographs had been made. Of these participants, only six patients were found to have UAPA, of whom three patients had no other cardiac anomalies. This means that one isolated UAPA was found among 200,000 men.

Recurrent pulmonary infections, decreased exercise tolerance, and mild dyspnea during exertion are the most common symptoms. Our finding that 37% of the patients had pulmonary infections is comparable with the results of earlier reports (39%).<sup>27</sup> Although the infections are usually mild, they may have devastating effects, such as necrotizing bronchopneumonia, leading to neonatal pneumonectomy.<sup>51</sup> In some patients, the frequent respiratory infections may be explained by bronchiectasis.<sup>16,17</sup> It has been suggested that alveolar hypocapnia can cause bronchoconstriction, while impaired mucociliary clearance and the diminished delivery of appropriate inflammatory cells may contribute to the high incidence of infections in patients with UAPA.<sup>16</sup>

Less frequently found are hemoptysis and signs of pulmonary hypertension. We found hemoptysis as the presenting symptom in 20% of the patients, which is comparable to the 18% reported earlier by Shakibi et al.<sup>27</sup>

Hemoptysis in patients with UAPA is caused by excessive collateral circulation.<sup>50</sup> The systemic collaterals to the

**Table 1—Summary of the Clinical Findings, Diagnostic Procedures, and Therapies Performed\***

Procedures	Present	Absent	Not Recorded
̇V/̇Q scan	55/94 (59)	39/94 (41)	14
Echo	34/100 (34)	66/100 (66)	8
HC	97/108 (90)	11/108 (10)	0
Inf	35/94 (37)	59/94 (63)	14
Dys	36/92 (40)	56/92 (60)	16
Hem	18/92 (20)	74/92 (80)	16
PHT	25/57 (44)	32/57 (56)	51
HAPE	7 (7)	101	
Pneu	9 (8)		
Rev	8 (7)		
Died	7 (7)		

\*Values given as No. of patients/total No. of patients (%).

̇V/̇Q = ventilation perfusion; Echo = echocardiography; HC = cardiac catheterization; Inf = recurrent respiratory infections; Dys = dyspnea at rest or diminished exercise capacity; Hem = hemoptysis; PHT = pulmonary hypertension; Pneu = pneumonectomy performed; Rev = revascularization of hilar arteries performed.

**Table 2—Reasons for Revascularization or Pneumonectomy**

Patient No./Reference No./Age	Description
1/54/29 yr	Massive hemoptysis for which left pneumonectomy was performed
2/51/23 d	Necrotizing bronchopneumonia for which right pneumonectomy was performed
3/30/3 yr	Recurrent hemoptysis; ligation and embolotherapy of arteriovenous malformations; eventually successful left pneumonectomy
4/25/21 mo	Severe pulmonary hypertension and congestive heart failure; significant improvement after revascularization of the right lung
5/2/9 yr	Revascularization of a right pulmonary artery
6/2/1 yr	Revascularization of a right pulmonary artery
7/24/3 mo	Pulmonary hypertension; restoration of normal pulmonary pressures after revascularization of the right pulmonary artery
8/66/3 mo	Cardiac failure and cyanosis; good clinical condition after revascularization of the right pulmonary artery
9/68/22 mo	Cardiac failure, cyanosis and pulmonary hypertension; disappearance of symptoms after reconstruction of right pulmonary artery
10/71/10 yr	Right pneumonectomy because of inflammatory processes and pulmonary hypertension; favorable outcome
11/20/5 yr	Right pneumonectomy because of recurrent bronchopneumonias; good clinical condition with 12 years follow-up
12/20/14 mo	Right pneumonectomy because of recurrent bronchopneumonias; good clinical condition with 2 years follow-up
13/39/28 yr	Chronic hemoptysis and pulmonary hypertension; revascularization of the right pulmonary artery; uneventful postoperative course
14/77/12 yr	Bronchiectasis in several lobes; right pneumonectomy
15/77/12 yr	Bronchiectasis; left inferior lobectomy
16/77/4.5 yr	Bronchiectasis; left inferior lobectomy
17/79/2 mo	Cardiac failure, cyanosis, pulmonary hypertension; successful restoration of right pulmonary vascularization

affected lung in patients with isolated UAPA come from the bronchial, intercostal, subclavian, or subdiaphragmatic arteries.<sup>52</sup> Before the development of the pulmonary artery, the lung is supplied by small transitory branches of the dorsal aorta that later disappear. These branches persist as enlarged supradiaphragmatic and infradiaphragmatic aortic branches, or as bronchial arteries in cases in which the pulmonary artery does not develop.<sup>1,34,52</sup> Hemoptysis may be self-limiting for many years<sup>35,52</sup> but also can lead to massive pulmonary hemorrhage and death.<sup>40,41</sup>

In the present review, in 25 of the 57 patients (44%) in whom pulmonary arterial pressure was measured, the pressure had increased. However, when related to the total group of 108 patients, the percentage of patients with pulmonary hypertension was comparable to those found previously (20 to 25%).<sup>27,28</sup> Which factors determine the presence or absence of pulmonary hypertension in patients with UAPA remain to be elucidated. Pulmonary hypertension in patients with UAPA also may be unmasked by predisposing factors such as HAPE and pregnancy.<sup>37–39,45–47</sup> When HAPE develops at modest altitudes (*ie*, < 3,000 m) predisposing factors are usually present.<sup>45</sup> Why some patients are prone to HAPE remains to be elucidated.<sup>81,82</sup> Some patients have pulmonary hypertension before the onset of pulmonary edema, and the combination of exercise and hypoxia lead to further exacerbation and HAPE.<sup>45</sup> The reactivity of the pulmonary vascular bed to hypoxia differs between individuals who are susceptible to HAPE and those not subject to it.<sup>46</sup> This also may be the case in subjects with an absent pulmonary artery, in whom a mild degree of pulmonary hypertension may be present.<sup>45</sup>

Various diagnostic possibilities are available for patients with UAPA. As a first step, a chest radiograph is usually

performed. This may show an absent hilar shadow, a shrunken affected lung, and a shift of the mediastinal structures to the affected side.<sup>35</sup> In addition, the absence of the left or right pulmonary artery, ipsilateral grossly diminished pulmonary vascular markings, a small hemithorax and intercostal bone space, ipsilateral cardiac and mediastinal displacement, ipsilateral hemidiaphragm elevation, and contralateral lung hyperinflation<sup>52</sup> may be present. Subsequently, an echocardiogram may confirm the diagnosis and exclude other cardiovascular abnormalities. Pulmonary hypertension also can be diagnosed. In this review, in only a minority of the patients (34%) was echocardiography performed. However, from 1990 onward echocardiography as well as CT scanning and MRI were more extensively used in the diagnostic workup of patients with UAPA. Both MRI and high-resolution CT scanning have proven valuable in the evaluation of congenital heart defects.<sup>83,84</sup> In UAPA patients, the distal pulmonary arteries can be visualized by these techniques, while high-resolution CT scanning also can assess the presence of bronchiectasis in cases of recurrent pulmonary infections.<sup>16</sup> Ventilation-perfusion scintigraphy can elucidate the anatomy of the affected side. When revascularization is proposed, cardiac catheterization is necessary, including pulmonary venous wedge angiography to discover hilar arteries. In this review, catheterization was performed in 90% of the patients, but pulmonary venous wedge angiography was performed in only four patients. It is not clear why pulmonary venous wedge angiography was performed in so few patients. Previous studies<sup>85,86</sup> in patients with pulmonary atresia or the occlusion of one pulmonary artery showed that pulmonary venous wedge angiography is a relatively safe and effective tool with which to visualize hidden pulmonary arteries. In those

**Table 3—Reasons for Not Performing a Heart Catheterization**

Patient No./Reference No./Age, yr	Description
1/36/60	Complete diagnosis was made by $\dot{V}/\dot{Q}$ scan, echocardiography, and CT scan
2/57/21	Admitted to hospital with hemoptysis; diagnosis was made by $\dot{V}/\dot{Q}$ scan; further diagnostic studies were refused by the patient
3/45/26	HAPE; only $\dot{V}/\dot{Q}$ scan performed
4/45/21	Severe HAPE; already dead on hospital admission
5/17/7	Diagnosis suspected on routine chest radiograph; only $\dot{V}/\dot{Q}$ scan performed
6/50/39	Progressive dyspnea; diagnosis made by $\dot{V}/\dot{Q}$ scan and CT scan
7/34/42	Decompression illness related to scuba diving; diagnosis made by echocardiography and MRI
8/37/24	Dyspnea during pregnancy; diagnosis made by $\dot{V}/\dot{Q}$ scan and CT scan
9/47/5	HAPE; diagnosis made by echocardiography and $\dot{V}/\dot{Q}$ scan
10/78/31	No significant complaints; abnormal chest radiograph; diagnosis made by CT scan and MRI

patients who had undergone surgery, a good correlation with preoperative findings was reported.<sup>85,86</sup> However, the need for cardiac catheterization may decrease with the more widespread use of echocardiography, CT scanning, and MRI (Table 3).

When pulmonary hypertension is present in a patient with UAPA, their condition may be improved by revascularization of the side with the absent artery (Table 2). In most cases, there is an identifiable artery at the hilum that may be used for revascularization.<sup>87</sup> In anatomic postmortem studies<sup>87</sup> and in surgical studies, hilar arteries usually are found.<sup>2</sup> The effect of revascularization has been described. In one case,<sup>24</sup> a 3-month-old child with severe pulmonary hypertension had normal pulmonary artery pressure after anastomosis of the hidden pulmonary artery. The child's condition remained stable during 7 years of follow-up. Another report<sup>25</sup> described a 21-month-old child with pulmonary hypertension and congestive heart failure in whom a hilar artery was reconstructed with a conduit (GORE-TEX; W.L. Gore and Associates; Flagstaff, AZ), leading to the normalization of pulmonary pressure and improvement of congestive heart failure. This condition remained stable during a follow-up period of 2 years.

If revascularization is not possible, or when pulmonary hypertension does not improve, therapeutic measures such as those described for patients with primary pulmonary hypertension may be helpful. Both in patients with primary pulmonary hypertension and in those with pulmonary hypertension secondary to congenital heart disease, long-term vasodilator therapy may improve survival.<sup>88,89</sup> Orally administered calcium channel blockers and continuous IV infusion of prostacyclin have been applied. However, we found no patient with UAPA and pulmonary hypertension in whom one of these therapies was applied. The therapy for HAPE consists of transport to a lower level as soon as possible and supplemental oxygen or nitric oxide if available.<sup>51,52</sup> Hemoptysis may be self-limiting over many years,<sup>35,52</sup> but massive hemoptysis can be treated only by selective embolization of the systemic collaterals,<sup>53</sup> or even pneumonectomy of the affected side.<sup>30,54</sup>

Many patients with UAPA can remain asymptomatic for a long period. However, the development of a pulmonary hemorrhage or pulmonary hypertension may preclude long-term survival. We found an overall mortality rate of

7%. Pulmonary hypertension may have devastating effects, especially when it develops during pregnancy. In this review, one of the three patients with pulmonary hypertension during pregnancy died. The high mortality rate of patients with pulmonary hypertension during pregnancy frequently has been reported,<sup>90,91</sup> leading to the advice to prevent pregnancy or terminate pregnancy at an early stage in cases of previously known pulmonary hypertension.<sup>91</sup> If the pregnancy is too advanced, extensive evaluation and therapy may help to improve outcome. Therapy may include treatment with vasodilators such as nifedipine and continuous NO inhalation.<sup>92,93</sup>

#### CONCLUSION

Only a few patients with UAPA remain asymptomatic. UAPA may be suspected by the presence of recurrent respiratory infections, hemoptysis, or pulmonary hypertension. The diagnosis of UAPA can be made by chest radiography and echocardiography. Anatomic details and the presence of bronchiectasis can be discerned by CT scanning and MRI. The presence of hilar arteries can be demonstrated by pulmonary venous wedge angiography. If sufficiently large hilar arteries are found, revascularization may significantly improve outcome. In the case of large collateral arteries leading to pulmonary hypertension or hemoptysis, embolization techniques can be used. The early diagnosis of UAPA thus can prevent further deterioration and can prevent the potentially devastating effects of high altitude or pregnancy.

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*Chest* 2002;122:1471-1477

DOI: 10.1378/chest.122.4.1471

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