particularly in cases of invasive thymoma where complete resection is technically difficult and hazardous, as in our patient. Hence, chemotherapy was administered and followed up with radiotherapy. The chemotherapeutic regimen used in this patient is considered the current standard and was initiated after normal cardiac function was restored. The increased risk of cardiomyopathy in patients with myotonic dystrophy makes the use of potentially cardiotoxic drugs debatable, but the benefits in this case were believed to outweigh the risks. To further reduce cardiotoxicity, doxorubicin was administered as a continuous infusion over 4 days rather than as a bolus. A PET scan performed at the completion of all therapy showed no evidence of thymoma despite the presence of a residual mass (presumably fibrous tissue) measuring 2 × 3 cm on CT scan that has remained stable over 6 months.

Thymoma is best treated by complete surgical resection when feasible. Noninvasive thymoma (Masaoka stage I) is completely resected with a recurrence rate of only 1.5%. No adjuvant therapy is required. Invasive or malignant thymoma (Masaoka stages II, III, and IV) is more difficult to treat. Surgery is still the mainstay with total resection where possible. Radiotherapy and chemotherapy are very useful because thymoma is responsive to both modalities. Adjuvant radiotherapy after complete resection reduces recurrence from nearly 30% to < 10%. After partial resection, radiotherapy often leads to a complete response. Chemotherapy, especially in a neoadjuvant manner, has been employed when surgery is not feasible at diagnosis. Surgical resection and adjuvant radiotherapy follow. When surgery is not possible, a combination of chemotherapy and radiation gives better results than either modality alone.1 In a study of seven patients who achieved a complete response with neoadjuvant chemotherapy, there was no residual tumor at surgery in two patients.2 We were able to achieve a substantial response on the basis of a CT scan and a complete response on the basis of a PET scan. PET scanning in thymoma has been shown to be a sensitive test.3 Thus, although there is a reasonable chance that the residual mass may not contain tumor cells, follow-up has been short and only time will tell since recurrences occur even after 30 years. The number of patients with myotonic dystrophy and thymoma is too small to draw any firm conclusions regarding treatment. We feel that a combined modality approach with chemotherapy and radiation is a safe and reasonable treatment option for these patients.

Table 1—Patients With Thymoma and Myotonic Dystrophy

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Age, yr</th>
<th>Sex</th>
<th>Country</th>
<th>Therapy</th>
<th>Outcome</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>51</td>
<td>Male</td>
<td>Japan</td>
<td>Radiotherapy, 16 Gy</td>
<td>No response; dead in 4 mo</td>
<td>4 mo</td>
</tr>
<tr>
<td>2</td>
<td>46</td>
<td>Female</td>
<td>Japan</td>
<td>None</td>
<td>Died suddenly 4 mo later</td>
<td>4 mo</td>
</tr>
<tr>
<td>3</td>
<td>32</td>
<td>Female</td>
<td>Spain</td>
<td>Radiotherapy, 50 Gy</td>
<td>Complete response</td>
<td>4 yr</td>
</tr>
<tr>
<td>4</td>
<td>64</td>
<td>Female</td>
<td>United States</td>
<td>Unknown</td>
<td>Unknown</td>
<td>Unknown</td>
</tr>
<tr>
<td>5</td>
<td>32</td>
<td>Male</td>
<td>United States</td>
<td>Thymectomy</td>
<td>Complete response; postoperative complications: embolism, pneumonia</td>
<td>Several months</td>
</tr>
<tr>
<td>6</td>
<td>46</td>
<td>Female</td>
<td>United States</td>
<td>Chemotherapy and radiotherapy</td>
<td>Complete response</td>
<td>19 mo</td>
</tr>
</tbody>
</table>

REFERENCES


Anomalous Collateral from the Coronary Artery to the Affected Lung in a Case of Congenital Absence of the Left Pulmonary Artery*

Effect on Coronary Circulation

George E. Kochiadakis, MD, Stavros I. Chryssostomakos, MD, Nikos E. Igoumenidis, MD, Emmanuel I. Skalidis, MD; and Panos E. Vardas, MD, PhD

A case of congenital absence of the left pulmonary artery, in which perfusion of the affected lung was accomplished via an arterial shunt from the circumflex coronary artery, is discussed. Data from myocardial perfusion scintigraphy showed that myocardial perfusion was unaffected by the existence of the...
shunt, largely because the flow through the shunt occurred mainly during systole. 

(CHEST 2002: 121:2063–2066)

Key words: flow wire; pulmonary artery agenesis; shunt

Isolated congenital absence of a pulmonary artery is a rare cardiovascular malformation the clinical course of which is benign, meaning that it may not be diagnosed until the patient reaches an advanced age. The perfusion of the affected lung in the case of the isolated absence of a pulmonary artery usually occurs via the bronchial arteries.

In this report, we describe a case of an isolated congenital absence of the left pulmonary artery, which was diagnosed in a patient at an advanced age and in which the perfusion of the affected lung was accomplished via an arterial shunt from the circumflex coronary artery.

CASE REPORT

A 62-year-old white woman was admitted to the hospital because of progressively increasing dyspnea, over 1 week, due to community-acquired acute bronchitis. The patient had a history of mild arterial hypertension and was known to have chronic bronchitis. She had never exhibited chest pain or other symptoms of ischemic heart disease.

On hospital admission, a physical examination was performed, and it revealed an obese, kyphoscoliotic person in respiratory distress, with a temperature of 37.9°C, arterial pressure of 145/88 mm Hg, and a pulse rate around 100 beat/min. She had mild lower extremity edema, and lung auscultation revealed diffused bronchospasm. There was a 3/6-holosystolic murmur in the right precordial area, and the liver was palpable and mildly enlarged. A chest radiograph showed an increased cardiac silhouette, bronchiectasis, kyphoscoliosis, elevation of the left hemidiaphragm, the absence of a left pulmonary artery main branch, and a small left hemithorax. Sinus tachycardia and incomplete right bundle-branch block were noted on the ECG. The echocardiogram revealed dilation and concentric hypertrophy of the right ventricle, whereas the left ventricle was structurally and functionally normal. Indications of severe pulmonary hypertension were also present. The left pulmonary artery main branch was not visualized. No significant findings were noted as a result of a blood test, except for a mild increase in leukocyte count. Antibiotics, bronchodilation, and oxygen administration were added to her treatment medications (which consisted of an angiotensin-converting enzyme inhibitor, furosemide, and a calcium channel blocker), and the patient’s symptoms improved after 4 days. At the same time, an investigation for secondary causes of pulmonary hypertension was programmed.

Lung perfusion imaging precluded the possibility of a pulmonary embolism, and an MRI suggested the diagnosis of congenital left pulmonary artery absence. Right heart catheterization confirmed the latter diagnosis. Pulmonary artery pressure was 42/21 mm Hg. Left heart catheterization and coronary angiography revealed that...
the left lung perfusion occurred via an arterial shunt from the left circumflex coronary artery (Fig 1). Aortography showed no arterial supply to the left lung other than that from the coronary artery.

With the aid of a flow-wire during coronary angiography the blood flow through the shunt was estimated and was found to reach 180 mL/min, mainly during the systolic phase of the cardiac cycle (Fig 2). The effect of the shunt on myocardial perfusion appeared to be negligible, since 201Tl scintigraphy showed uniform perfusion of the myocardium.

**Discussion**

The unilateral absence of a pulmonary artery is a rare congenital anomaly that may occur in isolation or in association with other congenital cardiovascular abnormalities, most frequently with the tetralogy of Fallot.1–4 It is believed that in patients with the Fallot anomaly, the pathologic process that results in absent mediastinal pulmonary arteries occurs early in gestation, at a time when other primitive vessels still exist connecting the aorta and its branches with the pulmonary vasculature. These vessels persist after birth and function as collateral arteries for the affected lung. In such patients the collateral arteries may have very unusual extrapulmonary courses. Entering the lungs, they typically supply segmental pulmonary arteries. These collaterals may originate from the aorta or the brachiocephalic branches. The coronary origin also has been well-described.5

In patients with an isolated absence of the left pulmonary artery, collateral flow to the lung is quite different. The process whereby the mediastinal pulmonary artery flow is disrupted occurs quite late in gestation or even after birth. In these patients, there is usually a gradual enlargement of normally present systemic-to-pulmonary connections, most commonly the bronchial arteries. The collateral vessels described earlier do not usually exist.

The patient described here had an isolated absence of the left pulmonary artery, in which the perfusion of the affected lung occurred via a vessel that arose from a coronary artery. To our knowledge, this is only the third such case described in the international literature.6 The existence of such a vessel could, in theory, have a negative effect on myocardial perfusion occurring via a steal phenomenon and induce myocardial ischemia. However, in our patient no such effect was detected using 201Tl scintigraphy. This was due to the fact that the left-to-right flow through the shunt occurred mainly during the systolic phase of the cardiac cycle, rather than during the diastolic phase, when myocardial perfusion takes place.

In conclusion, in a patient with a congenital absence of a pulmonary artery the possibility should be considered that the affected lung is being perfused by a shunt arising from a coronary vessel. Given that in all three cases...
reported so far the shunt was not discovered until the patient was at an advanced age, and that there was no chest pain or other sign of ischemic heart disease, it appears that the existence of such a vessel does not affect myocardial perfusion and so does not in itself indicate surgical intervention.

REFERENCES


New Procedure: Bronchoscopic Endobronchial Sealing*

A New Mode of Managing Hemoptysis

Parthasarathi Bhattacharyya, MD, DNB, DM; Anjan Dutta, MBBS, DA; Ananta Narayan Samanta, MBBS; and Samrat Roy Chowdhury, MBBS

Six patients with hemoptysis were treated by endobronchial sealing, with n-butyl cyanoacrylate, of the bleeding segment or subsegment. There was an immediate arrest of bleeding without any recurrence for a mean follow-up period of 127 (± 67.17) days. Endobronchial sealing appears to be an effective method of managing hemoptysis.

(CHEST 2002; 121:2066–2069)

Key words: endobronchial sealing, fiberoptic bronchoscopy, hemoptysis, n-butyl cyanoacrylate

Hemoptysis is a common clinical problem.¹ The source of bleeding lies, usually, in the tracheobronchial tree and rarely in the nose, pharynx, or larynx.² Management of hemoptysis aims to stop the bleeding, replenish the blood loss, and treat the underlying etiology. Hemostasis is often difficult to achieve when the conservative treatment fails. Several bronchoscopic techniques, such as the bronchial tamponade, have been used to arrest bleeding.³ We have adopted a method of treating hemoptysis by selective placement of a catheter in the bleeding segment (or subsegment), with the help of a fiberoptic bronchoscope, and instillation of a sealant, n-butyl cyanoacrylate, a biocompatible glue that has been used successfully in several other hemostatic and sealing procedures.⁴

METHODS AND MATERIALS

Selection of Patients

Patients with prolonged (> 7 days) hemoptysis despite continued conservative therapy were selected for the endoscopic procedure. Proper informed consents were obtained. A total of six patients (4 were male, 2 were female), with an average (±SD) age of 56.1 (± 8.33) yr, were treated with the endoscopic procedure (Table 1). Each patient was clinically evaluated for the possible site and cause of bleeding, and the approximate amount of blood loss was determined. The baseline investigations, such as percentage hemoglobin, packed cell volume, total and differential leukocyte count, erythrocyte sedimentation rate, prothrombin time, partial thromboplastin time, platelet count, and chest roentgenograms were obtained in all of them. CT scan (thorax) was done in four patients. There was no coagulation problem in any of the patients. Sputum smears for acid-fast bacilli were negative in all the patients on at least three different occasions. Two patients (Table 1), who were diagnosed to have pulmonary tuberculosis from the radiologic appearances, showed good clinical response to antitubercular drugs. One patient with past history of treatment for tuberculosis had multiple patchy areas of fibrosis and bronchiectasis. In the other three patients, the etiologic diagnoses were not clear. Fine needle and/or transbronchial needle aspirations revealed no malignancy on cytological examinations in two patients. The aspiration smears did not reveal any infective agent on Gram and Ziehl-Neelsen staining. The patients, however, responded positively to antibiotics. There was no contraindication to bronchoscopy in any of the patients according to the guidelines of the American Thoracic Society.⁵

Fiberoptic bronchoscopy was done in the morning hours after an overnight fasting. After mild sedation (intramuscular injection of promethazine, 25 mg, with atropine, 0.6 mg; 30 min before the procedure), the nose, pharynx, and upper airways were sprayed with lidocaine (4% solution), and fiberoptic bronchoscopy was performed (Pentax FB 18 P bronchoscope; Pentax, Tokyo, Japan) through transnasal route. Intra-airway spray of lidocaine (2%) was used as necessary. Blood from the tracheobronchial wall was cleared by saline, and the bleeding segment/site was detected. This was further confirmed by asking the patient to cough, resulting in fresh bleeding. A polyethylene catheter with an outer diameter of 2 mm was passed through the bronchoscope channel to place it slowly into the bleeding segment (Figure 1). Thereafter, 0.5 mL n-butyl cyanoacrylate glue was injected through the catheter with a water column behind, which was just adequate to flush the glue into the targeted area. The catheter was withdrawn within a few seconds along with the bronchoscope. The scope was passed again after 2 to 3 min to confirm the absence of bleeding. The same procedure was repeated until the hemostasis was achieved. Fresh bleeding was noticed after removal of the clot in one patient, but the procedure was successfully performed at the same sitting after ensuring hemostasis with bronchoscopic wedging and local instillation of adrenaline solution.

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