Coronary Artery-to-Pulmonary Artery Collaterals in Chronic Thromboembolic Pulmonary Hypertension

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Bronchopulmonary collateral vasculature is a significant source of systemic blood flow in patients with chronic thromboembolic pulmonary hypertension (CTEPH). We present 2 cases of coronary artery-to-pulmonary artery (PA) collateral vessels, which have not been previously described in patients with CTEPH.

Case 1
A 40-year-old man with a history of deep venous thrombosis after parotidectomy for a benign lesion, recurrent strokes, and right-sided pulmonary embolism was evaluated for 5 years of progressive exertional dyspnea and suspected CTEPH. He had no known hereditary thrombophilia. Preoperative ventilation-perfusion scintigraphy demonstrated absent perfusion to the right lung and small defects in the left lower lobe (Figure 1). Pulmonary angiography confirmed occlusion of the proximal right PA with no distal flow (Figure 2; Movie I in the Data Supplement). His mean PA pressure was 35 mm Hg, pulmonary vascular resistance was 511 dyne/s per cm^5, and cardiac index was 2.3 L/min per m^2. Coronary angiography revealed a 4.6-mm diameter corkscrew vessel arising from the left circumflex artery and traveling to the right lung (Figure 3; Movie II in the Data Supplement). He had no significant coronary artery disease. The patient underwent bilateral pulmonary thromboendarterectomy (PTE; Figure 4), during which a small atrial septal defect was found and repaired. Collaterals were noted throughout the mediastinum, particularly around the aorta traversing toward the right lung and directly penetrating the right main PA. Final mean PA pressure was 38 mm Hg, pulmonary vascular resistance was 296 dyne/s per cm^2, and cardiac index was 3.0 L/min per m^2. Improvements were evident on subsequent ventilation-perfusion scans. His hospital course was complicated by a superficial sternal wound infection and pericardial effusion, but he was discharged on postoperative day 24.

Case 2
A 46-year-old man presented to an outside facility with hemoptysis after completing antibiotics for pneumonia. A ventilation-perfusion scan revealed absent perfusion to the entire left lung. He was diagnosed with a left-sided pulmonary embolism and initiated on warfarin. Five months later, he was evaluated at our center for PTE for suspected CTEPH. He had no family history of deep venous thrombosis/pulmonary embolism. A repeat ventilation-perfusion scan showed overall poor perfusion to the left lung, particularly the lower lobe and apex (Figure 5). Pulmonary angiography verified a stump-occluded left PA with trivial distal flow (Figure 6; Movie III in the Data Supplement). Mean PA pressure was 50 mm Hg, pulmonary vascular resistance was 914 dyne/s per cm^2, and cardiac index was 1.8 L/min per m^2. Coronary angiography revealed a 3.7-mm diameter vessel arising from the left circumflex artery and traversing toward the left lung (Figure 7; Movies IV and V in the Data Supplement). He had no significant coronary artery disease. He underwent bilateral PTE (Figure 8). No congenital heart defect was found. Postoperative mean PA pressure was 23 mm Hg, pulmonary vascular resistance was 200 dyne/s per cm^2, and cardiac index was 2.8 L/min per m^2. His hospital course was uneventful.

Discussion
CTEPH can cause severe right heart failure with high morbidity and mortality, but it is curable with PTE. Identifying physiological changes and factors that portend better surgical outcomes are essential in caring for these patients. Whereas bronchopulmonary collaterals are well described in CTEPH and can account for 30% of the systemic blood flow, coronary–pulmonary collaterals have not been previously reported, either in pulmonary angiography or computed tomographic angiography studies, the core diagnostic modalities in CTEPH. A study of these patients using multidetector row helical computed tomography identified inferior phrenic, intercostal, and internal mammary artery collaterals, but not coronary–pulmonary collaterals, perhaps because of inadequate sample size or limitations of contrast timing.

At University of California at San Diego Medical Center, coronary angiography is performed to rule out obstructive coronary artery disease before PTE in men aged >40 years, women >45 years, and in younger patients with significant risk factors. Coronary artery fistulas are identified in 0.05% to 0.25% of patients undergoing coronary angiography; 15%...
to 30% are coronary–pulmonary. Most are asymptomatic. Limitations of this technique include contrast dilution, flow limitation, and 2-dimensional viewing. Coronary computed tomographic angiography may better demonstrate these fistulas; the incidence was 0.32% in 1 study.

The coronary–pulmonary collaterals in the 2 cases described here are unlikely to be incidental findings. These fistulas have been identified in pathologies such as Takayasu arteritis with severe pulmonary hypertension and pulmonary atresia with ventricular septal defect. These cases suggest that these vessels help maintain pulmonary circulation. The angiogenic consequences of unilateral PA obstruction on the bronchial system, but not the coronary system, have been examined in animal models, demonstrating increased bronchial artery diameter, length, and tortuosity. Blood flow may augment as much as a 300% in <2 weeks. In patients with CTEPH, this dilation and tortuosity persist at least 1 year after successful PTE and normalized PA pressure.

Bronchopulmonary collaterals are also correlated with significantly better postoperative mortality rates and hemodynamic response in patients with CTEPH. Coronary–pulmonary collaterals may denote proximal (and therefore operable) disease and carry similar prognostic value, because both these patients had significant hemodynamic improvement after PTE. CTEPH pathophysiology has not been fully elucidated but results from pulmonary vessel obstruction and subsequent small-vessel arteriopathy. One theory is that patients with proximal lesions may have less distal arteriopathy. The greater pressure difference between systemic circulation compared with pulmonary circulation distal to the occlusions may stimulate collateralization and explain better surgical outcomes.

The factors that promote coronary–pulmonary collateralization in CTEPH are unknown. These vessels likely help maintain pulmonary circulation. The prognostic implications of these findings require further study.

Disclosures

None.

References


Key Words: collateral circulation ◼ coronary circulation ◼ hypertension, pulmonary ◼ pulmonary circulation

Figure 1. In case 1, anterior and posterior views of the perfusion portion of the ventilation–perfusion scan highlighted no perfusion to the right lung and small defects in the left lower lobe.
Figure 2. In case 1, pulmonary angiography revealed occlusion of the proximal right pulmonary artery (frontal view).

Figure 3. In case 1, left coronary angiogram demonstrated a 4.6-mm diameter corkscrew collateral vessel arising from the left circumflex artery and emptying into the right bronchial territory. A, Right anterior oblique caudal projection. B, Left anterior oblique cranial projection.

Figure 4. In case 1, fibrous tissue measuring 6.0×4.8×1.4 cm in aggregate was removed from the right pulmonary artery (left) during pulmonary endarterectomy. Fragments were also removed from the left pulmonary artery (right).
Figure 5. In case 2, ventilation–perfusion scintigraphy highlighted poor perfusion to the left lung with large defects in the left lower lobe and left apex.

Figure 6. In case 2, pulmonary angiography confirmed an essentially stump-occluded left pulmonary artery (left lateral view).

Figure 7. In case 2, left coronary angiogram revealed large tortuous vessels traversing to the left lung. A, Right anterior oblique cranial view. B, Left anterior oblique cranial view.
Figure 8. In case 2, bilateral pulmonary endarterectomy resulted in removal of organizing thrombus measuring 5.2×3.5×0.7 cm from the right pulmonary artery (left) and 4.5×2.5×1.7 cm from the left pulmonary artery (right).
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Movie Legends

**Movie 1.** Pulmonary angiogram revealing occlusion of the proximal right pulmonary artery in case 1.

**Movie 2.** Left coronary angiogram in case 1 demonstrating a 4.6-mm-diameter corkscrew collateral vessel arising from the left circumflex artery and traveling to the right lung (depicted in the right anterior oblique caudal projection).

**Movie 3.** Pulmonary angiogram revealing occlusion of the proximal left pulmonary artery in case 2.

**Movie 4.** Left coronary angiogram in case 2 demonstrating a 3.7-mm-diameter collateral vessel arising from the left circumflex artery and traveling towards the left lung (depicted in the right anterior oblique cranial projection).

**Movie 5.** Left coronary angiogram in case 2 demonstrating a 3.7-mm-diameter collateral vessel arising from the left circumflex artery and traveling towards the left lung (depicted in the left anterior oblique caudal projection).