# Percutaneous Revascularization for Left Main Coronary Artery Compression From Pulmonary Artery Enlargement Due to Pulmonary Hypertension

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Extrinsic compression due to pulmonary artery enlargement from severe pulmonary hypertension is an uncommon cause of hemodynamically significant left main artery stenosis. Patients with severe pulmonary hypertension who experience angina should be evaluated for possible extrinsic compression of the left main artery due to pulmonary artery enlargement. Although computed tomographic angiography and cardiac magnetic resonance imaging are helpful in the screening for extrinsic left main artery compression, coronary angiography is the gold standard for the diagnosis. Percutaneous coronary intervention of the left main artery is feasible, safe, and a reasonable initial revascularization strategy for these patients because of the high risk of postoperative right ventricular failure and mortality observed with bypass surgery.

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**KEY WORDS**

Left main coronary artery • Pulmonary hypertension • Percutaneous coronary intervention

The vast majority of left main coronary artery disease, observed in 3% to 5% of patients undergoing coronary angiography, is due to atherosclerosis.[1,2] However, an uncommon cause of left main disease is extrinsic compression due to pulmonary artery (PA) enlargement secondary to severe pulmonary hypertension (PH). We present a case of a patient with severe PH leading to PA enlargement causing extrinsic left main compression and discuss different treatment options for this condition.

A 46-year-old woman with a past medical history of a large secundum atrial septal defect and PH was referred for cardiac catheterization and coronary angiography after multidetector computed tomographic (CT) coronary angiography demonstrated a high-grade eccentric compression of the proximal left main artery due to a PA aneurysm. Coronary angiography confirmed a severe stenosis of the left main artery. After discussion with the patient, the adult congenital heart disease cardiologist, and cardiac surgeon, the patient was referred for percutaneous coronary intervention (PCI) of the left main artery.

After pretreatment with aspirin, 325 mg, and clopidogrel, 600 mg, a 3.5 × 15 mm Xience V® (Abbott Vascular, Abbott Park, IL) was deployed in the left main artery at 16 atmospheres. The left main artery was postdilated with a 3.75 × 12 mm noncompliant balloon. Intravascular ultrasound (IVUS) demonstrated optimal expansion and apposition of the stent and confirmed coverage of the ostium of the left main artery. Follow-up CT angiography demonstrated a widely patent stent, and the patient continues to do well with no clinical evidence of ischemia.

The exact incidence of extrinsic left main artery compression due to PA enlargement is unknown, but ranges from 5% to 44% of patients with PH.[3-5] Although no clear risk factors have been identified for the development of extrinsic left main artery compression, case reports suggest that it occurs more often in patients with longstanding and severe PH. The pathogenesis of extrinsic left main artery compression is due to PA enlargement from PH.[6] The proximity of the left main artery to the PA and the compression by the fascial sheath surrounding the great vessels make the left main artery susceptible to extrinsic compression by an enlarged PA.[6,7]

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PH observed in patients with extrinsic left main artery compression is most frequently due to congenital heart disease (observed in 83% of patients), with atrial septal defects being the most common cause.[4] Other etiologies include idiopathic PH, chronic thromboembolic pulmonary hypertension, and advanced parenchymal lung disease.[7-10] Uncommon causes of extrinsic left main artery compression include thoracic outlet syndrome, syphilis, metastatic squamous cell carcinoma, and bronchogenic cyst.[2,11,12]

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Vascular remodeling that occurs gradually appears to be required as acute increase in the intraluminal pressure of a normal PA does not lead to PA dilatation and extrinsic compression of the left main artery.[13] Chronic severe PH in the proximal pulmonary vessels can lead to intimal and medial hypertrophy, fibrosis, luminal dilatation, and atherosclerotic plaque formation.[14] PA dilatation with a mean PA trunk diameter of 55 mm and a mean main PA to aortic root diameter ratio of 2.0 is typically seen.[4,8] PH is not required for PA dilatation and subsequent compression of the left main artery.[10] The left main artery is displaced inferiorly and in close proximity with the left aortic sinus due to the PA dilatation.[8]

Clinicians should have a high index of suspicion of extrinsic left main artery compression from PA enlargement for patients with severe PH who experience chest discomfort or dyspnea. These patients may not have risk factors for coronary artery disease and may not undergo evaluation for myocardial ischemia with coronary angiography. Approximately 40% of patients with PH experience chest pain that resembles angina.[14] Chest pain may be due to right ventricular strain and ischemia, PA dilatation, and a reduction in the coronary perfusion gradient due to elevated right atrial pressures.[14-17] Patients may also present with angina-equivalent symptoms such as syncope,[18,19] myocardial infarction,[20,21] left ventricular dysfunction/cardiogenic shock,[20,22] malignant arrhythmias, and sudden death.[3,23]

The gold standard for the diagnosis of extrinsic left main artery compression is coronary angiography. The left main artery typically narrows eccentrically at the

ostium, tapers open at the distal end, and is often well visualized at a 45° left anterior oblique view with a 30° cranial angulation.[4,8,24,25] There is also inferior displacement of the left main artery, creating an acute angle between the artery and the left coronary cusp.[24] IVUS commonly reveals a slit-like ostial left main stenosis as a result of extrinsic compression that is devoid of significant atherosclerotic plaque.[21,22,26,27] Fractional flow reserve can be performed to determine the severity of the left main artery stenosis.[22,27,28]

Although the sensitivity is unknown, multidetector CT coronary angiography and cardiac magnetic resonance imaging (MRI) are commonly used noninvasive imaging modalities to detect extrinsic left main artery compression from PA enlargement. Multidetector CT coronary angiography can reveal extrinsic compression of the left main artery, the luminal diameter of the left main artery, assessment of the PA trunk diameter, and the angle of the takeoff of the left main artery from the aortic root.[2,20,28-30] Cardiac MRI can also demonstrate extrinsic left main artery compression from PA enlargement as well as assess ventricular function and structure.[10,28] It is at the physician's discretion to choose CT or MRI on a case-by-case scenario as there are no comparative data with the two imaging modalities. Several factors should be considered when choosing CT angiography over cardiac MRI. CT angiography may be preferred in patients with implanted hardware (eg, pacemaker, defibrillator, cerebral aneurysm clips, insulin pumps), claustrophobia, and inability to hold their breath and remain prone for a longer period of time. Cardiac MRI may be preferred in patients with iodine-based contrast allergy or chronic renal insufficiency. However, patients on hemodialysis should not receive gadolinium-DTPA because of the risk of systemic nephrogenic fibrosis. Centers may have personnel that specialize in performing and interpreting one particular imaging modality over the other and may be the preferred study. Both modalities are contraindicated in pregnant women.

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Nuclear myocardial perfusion imaging has relatively low sensitivity and therefore is not reliable in the diagnosis of extrinsic left main artery compression due to PA enlargement from severe PH. Nuclear perfusion imaging detected regional ischemia in only four out of 10 patients with extrinsic left main artery compression.[4,24,31,32] Echocardiography may reveal a PA trunk diameter greater than 40 mm and PA to aortic root diameter ratio greater than 1.21 in at-risk patients.[4] However, it is inadequate for the diagnosis of extrinsic left main artery compression.

Treatment options for extrinsic left main artery compression include vasodilator therapy,[18,22] percutaneous and surgical revascularization, correction of the underlying congenital heart defect, thromboendarterectomy, and lung or heart-lung transplantation.

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The 2009 American College of Cardiology/American Heart Association/Society for Cardiovascular Angiography and Interventions (ACC/AHA/SCAI) focused guidelines for PCI updated the class of recommendation of PCI for patients with unprotected left main PCI from class III to class IIb. PCI may be considered in patients with anatomic conditions associated with a low risk of procedural complications and clinical conditions that predict an increased risk of adverse surgical outcomes.[33] The 2010 joint European Society of Cardiology and European Association for Cardio-Thoracic Surgery guidelines on myocardial revascularization state that unprotected left main coronary artery PCI involving the ostium/midshaft with or without single vessel coronary disease is a class IIa recommendation.[34] Given that patients with extrinsic left main artery compression with severe PH fall within these categories because they are at high risk of adverse surgical outcomes and the location of the compression is at the ostium of the left main artery, PCI appears to be the preferred revascularization strategy.[9] PCI is technically feasible, can be performed with one stent, is associated with excellent angiographic success, and is likely to be associated with low restenosis and thrombosis rates because of the lack of significant atherosclerotic burden at the ostial location. The left anterior oblique-cranial view frequently provides optimal visualization during placement of the stent at the left main ostium. IVUS provides confirmation of adequate ostial coverage by the stent and optimal stent

expansion and apposition. Although long-term data are limited in these patients, they demonstrate the sustained safety and efficacy with elective unprotected left main PCI in atherosclerotic lesions.[35,36]

Both drug-eluting stents and bare-metal stents are viable treatment options; the risk of restenosis is likely to be low given the ostial lesion location, the large diameter of the left main artery, and the fact that the underlying pathology does not involve atherosclerosis in this patient population. In ostial or midshaft left main disease secondary to atherosclerosis, the rate of target lesion revascularization after PCI with drug-eluting stents was remarkably low (0.7% at a mean follow-up of ~2.5 years).[35] The rates of definite and probable stent thrombosis for elective PCI of the left main artery with drug-eluting stents were also very low (0.9% at a mean follow-up of nearly 2.5 years).[37] In patients with no contraindication to aspirin and clopidogrel for a minimum duration of 1 year, PCI with drug-eluting stents should be considered as the first choice of therapy, given the potential for sudden death in patients who had in-stent restenosis of the left main artery.[9]

The ACC/AHA/SCAI 2009 focused PCI guidelines do not recommend routine follow-up angiography as no study has demonstrated its clinical benefit.[33] Furthermore, the performance of follow-up angiography may lead to unnecessary repeat revascularization, especially if fractional flow reserve is not performed.

Surgical revascularization is uncommonly performed for extrinsic left main artery compression because of the high surgical mortality associated with PH.[38] In contrast, percutaneous revascularization is not associated with the increased risk of postoperative right ventricular failure and mortality, which may occur after bypass surgery. The data on stand-alone bypass surgery with the use of a left internal mammary artery grafting to the left anterior descending artery for the treatment of left main artery compression are limited.[4] Surgical revascularization with concomitant pulmonary thromboendarterectomy,[10] correction of underlying congenital heart defect,[7] or PA reconstruction[31] have been reported. Pulmonary thromboendarterectomy[10,24,26] or repair of the atrial septal defect[2,6,30] may decrease PA pressures and size of the aneurysm and therefore reduce the extrinsic compression of the left main artery.[7] Therefore, surgical revascularization may be reserved for those who need concomitant corrective surgical repair of the underlying pathology, as PCI is a recognized treatment for these high-risk patients. The only definitive therapy for severe PH and extrinsic left main compression is lung or heart-lung transplantation.[4,25] PCI may serve as a bridge until transplantation is performed.[39]

## Conclusions

Extrinsic compression of the left main artery can be caused by PA enlargement due to severe PH and should be considered in patients with PH and angina and left ventricular dysfunction. Chronic severe PH can lead to PA remodeling and dilatation, which leads to the inferior displacement and compression of the left main artery. CT angiography or cardiac MRI may provide a preliminary diagnosis of extrinsic left main artery compression, which can be confirmed with invasive coronary angiography. Percutaneous revascularization is a safe, technically feasible, effective treatment strategy, and should be recognized as the preferred revascularization strategy in those patients who are poor surgical revascularization candidates due to the high risk of postoperative right ventricular failure and mortality due to severe PH. Though a randomized clinical trial comparing the two revascularization strategies would be ideal, it is unlikely to be conducted because of patient preference for the less invasive revascularization strategy, logistical complexities of such a large undertaking, and the rarity of the condition. ■

*Surgical revascularization is uncommonly performed for extrinsic left main artery compression because of the high surgical mortality associated with PH.*

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## MAIN POINTS

- An uncommon cause of significant left main stenosis is extrinsic compression due to pulmonary artery enlargement from severe pulmonary hypertension.
- Patients with severe pulmonary hypertension who experience angina should be screened for possible extrinsic compression of the left main artery due to pulmonary artery enlargement with computed tomography angiography and cardiac magnetic resonance imaging.
- Coronary angiography is the gold standard for the diagnosis of left main artery compression.
- Stenting of the main artery is a feasible, safe, and viable revascularization strategy for these patients because bypass surgery is associated with a high risk of postoperative right ventricular failure and mortality.