

Extrinsic Compression of the Left Coronary Ostium by the Pulmonary Trunk

Management in a Case of Eisenmenger Syndrome

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Extrinsic compression of the left main coronary artery by a massively dilated pulmonary artery in patients who have severe pulmonary hypertension can lead to significant myocardial ischemia. A 58-year-old man with a large patent ductus arteriosus and Eisenmenger syndrome presented with angina at rest and worsening heart failure of 3 months' duration. The new symptoms were recognized to be secondary to extrinsic compression of the left main coronary artery ostium by a dilated main pulmonary artery and were successfully relieved by the placement of a metallic stent in the affected segment of the left main coronary artery. Multislice computed tomographic imaging after 6 months showed stent patency and the intimate relation of the stented vessel to the dilated main pulmonary trunk. We discuss diagnostic and management issues pertaining to this uncommon clinical entity. (Tex Heart Inst J 2010;37(1):95-8)

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A pulmonary trunk dilated by severe pulmonary artery hypertension (PAH) can compress the aortic ostium of the left main coronary artery (LMCA).¹ If the primary disease process causing PAH cannot be reversed, progressive coronary narrowing can lead to left ventricular myocardial dysfunction, angina, and worsening of heart failure.² We report the case of a 58-year-old patient with a large patent ductus arteriosus and Eisenmenger syndrome who presented with angina at rest and worsening of heart failure due to left ventricular dysfunction. Coronary angiography showed extrinsic compression of the LMCA, at its origin, by a dilated pulmonary trunk. Stenting of the LMCA resulted in immediate relief of angina and led to symptomatic improvement.

Case Report

In February 2008, a 58-year-old man presented at our hospital. In his early adolescence, he had undergone cardiac catheterization, which had yielded a diagnosis of large patent ductus arteriosus with inoperable pulmonary vascular resistance. During the 3 months before the 2008 presentation, the patient developed bipedal edema, increasing effort intolerance, and severe chest pains at rest that radiated to the neck and jaws. His atherosclerotic risk factors included noninsulin-dependent diabetes mellitus and systemic hypertension (both present for 5 years) and hyperuricemia secondary to polycythemia. On physical examination, he had polycythemia, differential cyanosis and clubbing (the oxygen saturation in the toes was 78% on pulse oximetry), elevated jugular venous pressure with prominent A waves, cardiomegaly, visible pulmonary artery pulsations in the left 2nd intercostal space, and a closely split 2nd heart sound with a loud pulmonary component. Echocardiography revealed biventricular hypertrophy, mildly impaired left ventricular systolic function, a 20-mm ductus arteriosus with right-to-left flow, dilated pulmonary arteries, and competent tricuspid and mitral valves.

Coronary angiography was performed, after informed consent, to evaluate the patient's anginal episodes in consideration of his atherosclerotic risk factors. Catheterization showed severe PAH, with pulmonary vascular resistance of 19.2 Wood units. Severe smooth narrowing of the LMCA ostium was noted on cranially angulated projections, the distal LMCA measured 4 mm, and the branches were free of atherosclerotic irregularities (Fig. 1A). Caudal views upon left coronary injection did not show the ostial narrowing of the LMCA, which indicated the possibility of extrinsic uni-

directional compression by the dilated pulmonary artery. Right coronary injection showed septal branches of the left anterior descending coronary artery filling through collateral vessels; these were also free of irregularities.

Stenting of the LMCA ostium was done with the intention to provide a metallic scaffold of support against the extrinsic compression. A 0.014-inch floppy-tipped coronary guidewire—passed through a 6F EBU (Extra Backup) coronary guiding catheter—was parked in the left anterior descending coronary artery, and the left coronary ostium was stented with a 13-mm-long Bx Velocity™ RX stent (Cordis Corporation, a Johnson & Johnson company; Miami Lakes, Fla) to a diameter of 4 mm without any hemodynamic disturbances (Fig. 1B). This bare-metal 316L stainless-steel stent, with a closed-cell Flex Segment design, has good radial resistance to the stress of extrinsic compression of the pulmonary artery at systemic blood pressures.³ Dual antiplatelet therapy, optimal control of diabetes and hypertension, and decongestive therapy for heart failure were continued. The patient experienced immediate relief of anginal symptoms. At his 6-month follow-up visit, a multislice computed tomographic study with contrast showed a widely patent stent in the LMCA in proximity to the dilated main pulmonary artery (Figs. 2 and 3).

Discussion

Chest pain in patients who have severe PAH may be due to subendocardial right ventricular ischemia, to a reduced coronary perfusion pressure gradient secondary to elevated right atrial pressures, or sometimes to painful distention of the pulmonary trunk.⁴ It may also occur secondary to coronary atherosclerotic narrowing. Left ventricular dysfunction is observed in about 20% of patients who have severe PAH due to ventricular interdependence or myocardial ischemia.⁵

A dilated hypertensive main pulmonary artery that compresses the LMCA can lead to myocardial ischemia and sudden cardiac death.¹ Because the extrinsic coronary compression is functional, postmortem collapse of the pulmonary artery results in an underestimation of this compression at autopsy.⁶ Patients who have severe PAH but no atherosclerotic risk factors rarely undergo coronary angiography; hence, antemortem diagnosis is seldom made. Extrinsic compression of the left coronary ostium occurs in a single plane and is best profiled in cranial views; angiographic screening performed with a single injection of contrast medium in a caudal view can miss the diagnosis entirely.⁷

Anecdotally, LMCA ostial compression as a cause of angina or ischemic left ventricular dysfunction in patients with severe PAH is correctable.⁸ Such compression has been observed when the main pulmonary artery diameter exceeds 40 mm.⁹ Because Eisenmenger pa-

tients have so little effort tolerance, stress studies to document myocardial ischemia objectively may be difficult to perform. Once anatomic detection has been accomplished, the angiographic severity of the stenosis establishes the basis of treatment. The anatomic and hemodynamic significance of extrinsic narrowing of the LMCA in severe PAH has been analyzed by means of combined intravascular ultrasonography and pressure-

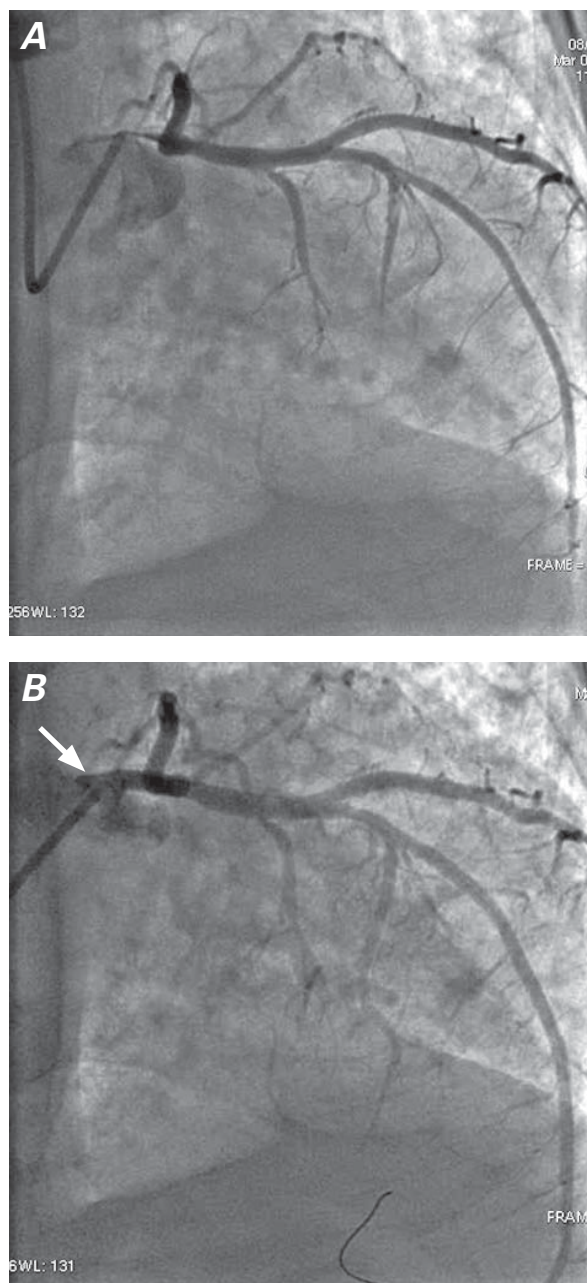


Fig. 1 A) Before stenting: note severe ostial narrowing of the left main coronary artery on selective coronary angiography, in a cranially angulated projection. **B)** After stenting: note wide lumen (arrow) of the left main coronary artery's ostium.

Real-time motion images are available at www.texasheart.org/journal.

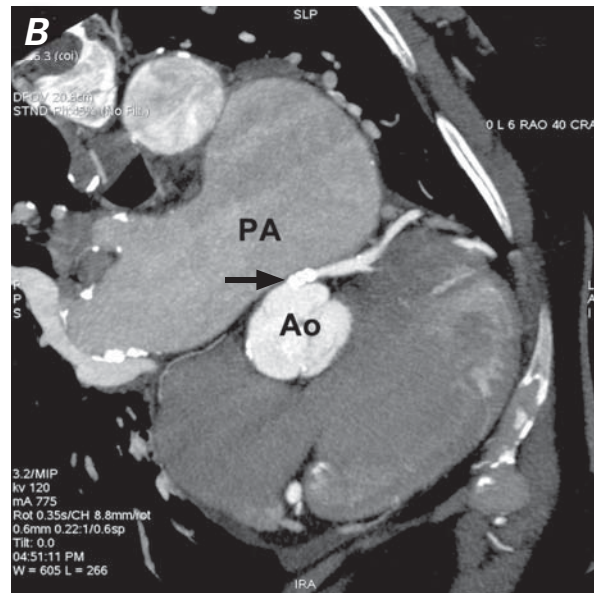
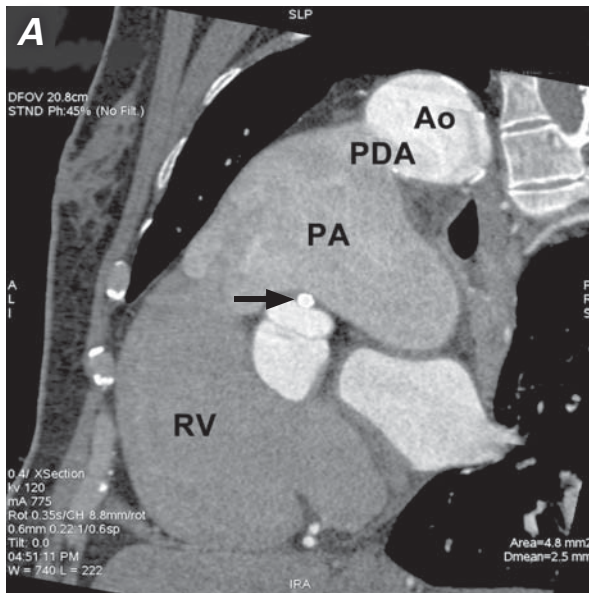


Fig. 2 Maximum-intensity projection from 64-slice computed tomographic image performed 6 months after stenting procedure. **A)** The relation of the stent (arrow) in the left coronary artery to the dilated pulmonary artery (PA). **B)** Another view of the massively dilated PA; the left main coronary artery stent (arrow) lies immediately below the PA.

Ao = aortic root; PDA = patent ductus arteriosus; RV = right ventricle

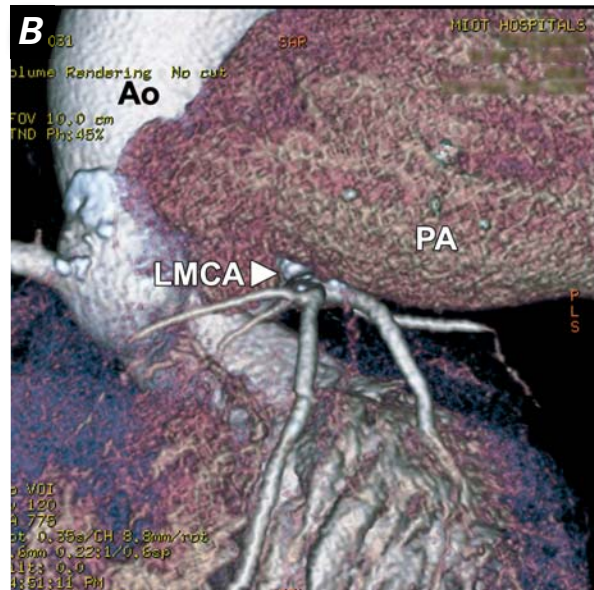
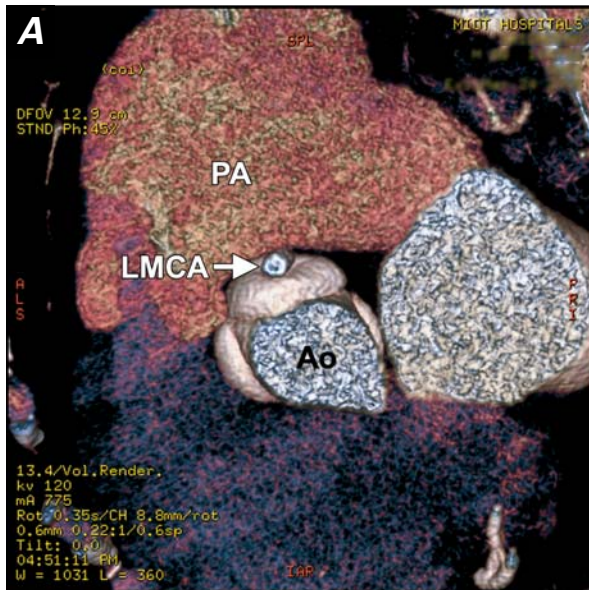


Fig. 3 Three-dimensional volume-rendered computed tomographic images. **A)** Arrow points to the stent in the left main coronary artery (LMCA) where it arises from the aortic root. Note the relationship of the stented LMCA to the dilated pulmonary artery (PA). **B)** The LMCA (arrowhead) is seen branching to the left anterior descending and circumflex coronary arteries.

Ao = aortic root

wire studies.¹⁰ In these patients, surgical myocardial revascularization carries a prohibitive risk of postoperative right ventricular failure.¹¹

As we have said above, we chose coronary stenting of the LMCA in order to protect—with a metallic scaffold—the lumen from extrinsic compression.^{3,12} Patients who have atherosclerotic endothelial dysfunction are pre-

disposed to severe in-stent restenosis, consequent to neointimal hyperplasia after stent implantation in coronary arteries. Although drug-eluting stents can prevent restenosis by inhibiting neointimal growth, long-term dual antiplatelet therapy is warranted to guard against catastrophic acute stent thrombosis in the left main coronary ostium.¹³ Patients with severe PAH are prone to pulmo-

nary apoplexy and hemoptysis, and long-term dual antiplatelet therapy puts them at a higher risk for such events.¹⁴ In patients who have Eisenmenger syndrome with hypoxia and cyanosis, atherosclerosis is relatively uncommon due to the hypocholesterolemia, up-regulated nitric oxide, and low platelet levels that are seen in hypoxia.

In the absence of atherosclerotic irregularities in the coronary arteries, coronary endothelial function in our patient with Eisenmenger syndrome is likely to be normal. Hence, we chose to deploy a bare-metal, non-drug-eluting, large (4-mm) 316L stainless-steel stent that has favorable radial stress-strain characteristics.³ This permits minimal endothelialization of the stent and guards against acute stent thrombosis at a later stage. In our patient, a 6-month follow-up angiographic study with multislice computed tomography showed good patency of the stent.

In summary, we describe a relatively uncommon clinical entity—extrinsic coronary compression leading to deterioration in a patient with Eisenmenger syndrome. Its diagnosis is solely dependent upon cranially angulated selective angiography, because the compression is uniplanar in nature.

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