**Video 4.** Angiography showed the prolongation of the dissection proximally and distally in the left anterior descending coronary artery

**Video 5.** Angiography showed the final result of the angioplasty of the left anterior descending coronary artery

**References**


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Left main coronary artery occlusion by external compression with a large pulmonary artery in Eisenmenger syndrome

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**Introduction**

The compression of the left main coronary artery (LMCA) with a dilated pulmonary artery (PA) is an important and life-threatening issue, possibly related to sudden death in patients with advanced pulmonary arterial hypertension (PAH). Here we present an exceptional and extreme case with an LMCA occlusion, who fortunately survived due to well-developed coronary collateral retrograde flow.

**Case Report**

A 39-year-old woman suffering from exertional dyspnea and frequent anginal episodes was referred to our center. PAH, secondary to unoperated patent ductus arteriosus, had been diagnosed 13 years before, and the patient was on a triple-combination therapy (bosentan, tadalafil, and inhaled iloprost). The clinical presentation involving cyanosis and clubbing jugular vein distension, loud S2P2, and parasternal lift was concordant with Eisenmenger’s syndrome. Electrocardiography indicated the right axis deviation, right ventricular strain pattern and anterolateral ischemia was suspected (Fig. 1a). Echocardiography revealed a dilated PA and right chambers with a leftward septal shift (Videos 1 and 2), and the estimated PA systolic pressure was 110 mm Hg. The LMCA compression was suspected. Computed tomography showed a complete LMCA occlusion due to external compression by the PA aneurysm (Fig. 1b and 1c). Cardiac catheterization revealed an advanced PAH (mean PA pressure, 88 mm Hg; pulmonary vascular resistance, 22.2 woods unit; cardiac index, 1.6 L/min/m²). Aortography and selective coronary angiography confirmed the LMCA occlusion (Fig. 1d and 1e, Videos 3 and 4). Coronary angiography also proved that the circumflex coronary artery originated from the right sinus of Valsalva, and the patient survived because of an efficient retrograde flow from the circumflex and right coronary arteries (Fig. 1f and 1g; Videos 5 and 6). Coronary by-pass surgery and PA aneurysm repair was offered. However, the patient refused surgery, and it was decided to switch the inhaled iloprost to parenteral prostanoid therapy.

**Discussion**

Sudden cardiac death is a common mode of mortality in patients with PAH, and it may be related with mechanical complications (1). The severe LMCA compression rate was 8.2% in a recent study and required intervention in most of the patients (2). The LMCA compression risk is related to the PA diameter. When the PA diameter exceeds 40 mm, it is defined as a PA aneurysm (3). PA aneurysms are mostly located in the main PA and can cause symptoms due to compression of adjacent tissues. Idiopathic, iatrogenic, infectious or connective tissue diseases, and vasculitis can cause a PA aneurysm, but the majority of the cases are related with congenital heart disease and PAH (3). As the LMCA compression can exist without anginal symptoms, physicians should be aware of the sudden death risk, and computed tomography should be performed to define the LMCA compression in patients at risk; particularly if the PA diameter is over 40 mm, ensuring the existence of a PA aneurysm (1, 2). Recent evidence indicates the coronary stenting as a viable option for resolution of LMCA compression (2). But in this case, a complete obstruction of LMCA was not amenable to stenting, so a surgical intervention was the only option. In addition to the mechanical compression of LMCA, considering the potentially mortal pulmonary rupture and dissection risk, our decision was also concordant with current recommendations for the management of PA aneurysms (3).

**Conclusion**

A marked dilatation of PA in patients with PAH may cause symptoms due to the compression of adjacent tissues, and particularly the LMCA compression may expose them to the risk of sudden
Video 6. The circumflex coronary artery was abnormally originating from the right sinus of Valsalva and selective angiography reveals collateral flow to the left coronary artery almost reaching to the left main ostium

References


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