

Discussion

The management of calcified coronary artery lesions remains one of the major limitations of PCI (1). IVL has recently emerged as a new technology that may efficaciously address calcified coronary or lower extremity atherosclerotic calcified lesions (2, 3). Via a balloon inflated in the vessel, IVL delivers pulsatile sonic pressure waves converted to mechanical energy that may modify vascular calcium by the induction of calcium fractures, allowing for proper stent expansion (4). Recently, a registry including 71 patients and a prospective study with 120 patients with calcified coronary lesions demonstrated the safety and effectiveness of the IVL technique (5, 6). In some case series, IVL has been used with success in the case of stent under-expansion resistant to conventional approaches (7, 8). Moreover, IVL was shown as a valuable option for modifying femoroiliac stenosis to facilitate delivery of a transfemoral Impella CP (9, 10).

Conclusion

We report the first case of LMT IVL supported by a microaxial pump assist device to facilitate stent expansion in a patient with porcelain aorta owing to congenital HDL deficiency.

Informed consent: The patient was informed, and she gave her consent for publishing this case report.

Video 1. LAO cranial coronary angiography performed via a 7-Fr guiding catheter demonstrating significant stenosis of the LMT artery. In addition, the massive calcifications of the ascending aorta (porcelain aorta) and, specifically, at the level of the LMT origin, are well visualized. A wire is advanced into the left anterior descending coronary artery. The Impella device is placed in the left ventricle.

Video 2. Fluoroscopy showing the advancement of the Impella device over a 0.018-inch wire from the abdominal into the ascending aorta.

Video 3. Left coronary system angiography after angioplasty showing the favorable result at the end of the procedure.

Video 4. Digital subtraction angiography of the right ilio-femoral axis showing femoral complete hemostasis with 2 Proglide suture-based closure devices after Impella sheath removal. The angiography is performed through a 0.035-inch over-the-wire peripheral balloon advanced crossover from the left femoral sheath and used as endovascular clamping at the time of sheath removal.

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An impressive image of unilateral pulmonary artery agenesis associated with coronary collateralization in an adult

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Introduction

Isolated unilateral absence of pulmonary artery (UAPA) is a very rare congenital anomaly in adults. The prevalence of this anomaly is about 1 in 200.000 live births. It is usually diagnosed

in early childhood and the average age of patients with the condition is 14 years (0–58 years) (1, 2). Clinical symptoms of the patient, anatomical variation of the pulmonary artery (PA), and additional cardiovascular anomalies together with their associated collateral circulation and pulmonary hypertension (PHT) are important factors to consider in the choice of treatment (3).

Case Report

A 63-year-old female was referred to our clinic for evaluation of progressively-worsening dyspnea over the last 1 year by a pulmonology clinic. We learned that she had exercise-induced dyspnea for 20 years. She was also hypertensive and had been diagnosed with atrial fibrillation for the past 2 years. Her physical examination and laboratory findings were within the normal range. An elevated right hemidiaphragm with hypoplasia of the right lung and a shift of the heart to the right side was seen on a routine chest X-ray. Her electrocardiogram revealed atrial fibrillation with normal ventricular rate response. Echocardiography revealed normal left ventricular systolic function and a normal left atrial diameter. We detected an increase in the systolic PA pressure (PAP=60 mm Hg) with normal right ventricular function. Lung scintigraphy showed a heterogeneous and decreased perfusion in the right hemithorax compared to the left side. We decided to perform computed tomographic pulmonary angiography (CTPA), since the chest X-ray was abnormal, to exclude the chronic thromboembolic pulmonary hypertension (CTEPH). CTPA revealed an absence of the right PA and dilated left and main PAs. Through magnetic resonance angiography of the aorta and PA, we confirmed the absence of the right PA and its branches, together with any additional anomalies. We performed right heart catheterization and coronary angiography to exclude coronary artery disease. The PAP as measured was 55/25 mm Hg with a mean value of 36 mm Hg and the pulmonary capillary wedge pressure was 15 mm Hg. Pulmonary angiography also showed an absence of the right PA with a dilated left PA and the vasoreactivity test was negative. Coronary angiography revealed normal coronary arteries, although we detected advanced collaterals arising from the conus branch of the right coronary artery, supplying the right PA's branches (Figs. 1, 2, Videos 1, 2). The patient was discharged with sildenafil and oral anticoagulant therapy. At the three-month follow-up, there was a significant improvement in the clinical condition. We did not replace her therapy with an endothelin receptor antagonist or riociguat.

Discussion

Cardiovascular anomalies such as septal defects, Fallot's tetralogy, patent ductus arteriosus, and right aortic arch may accompany UAPA (4). The isolated absence of the right PA is more common than that of the left (63%). Right-sided UAPA is less associated with other cardiac anomalies and patients with this

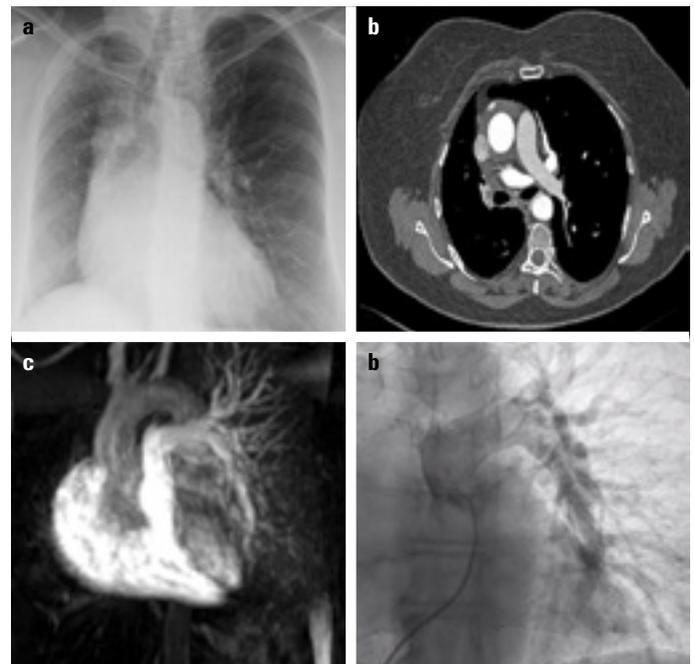


Figure 1. (a) Chest X-Ray demonstrates right hemithorax hypoplasia with an elevated right hemidiaphragm. (b) Computed tomography slice shows the absence of the right pulmonary artery. (c) Magnetic resonance angiography demonstrates the absence of a right pulmonary artery. (d) Pulmonary angiography shows the absence of the right pulmonary artery with a dilated left pulmonary artery

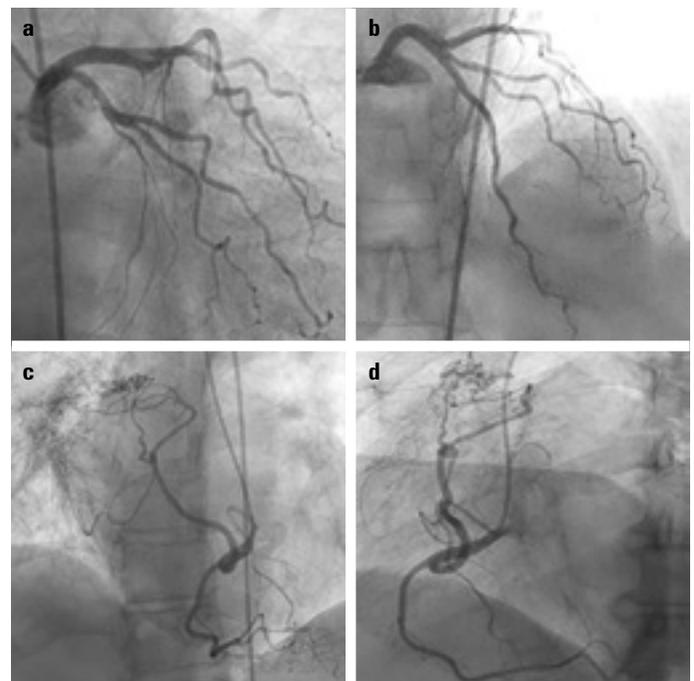


Figure 2. (a, b) Coronary angiography shows a normal left coronary arteries. (c, d) Right coronary angiography demonstrates a normal right coronary artery with collaterals from the conus branch to the right lung tissue

condition may survive to an advanced age (2). According to the literature, hemoptysis was present in 20%, recurrent pulmonary infections in 37%, limited exercise tolerance in 40%, and PHT in

44% of patients with isolated UAPA. The condition also affects both sexes equally (2). Echocardiography is useful for monitoring the right heart function and the development of PHT. Pulmonary angiography is the gold standard for the diagnosis of pulmonary angiography agenesis and the evaluation of collateral circulations (5). Another method that may be used in the differential diagnosis is magnetic resonance imaging, which is very valuable in terms of clearly showing the underlying etiology and comorbidities if it can be done routinely. Mortality is usually associated with progressive PHT and the resulting right ventricular failure, respiratory failure, and massive hemoptysis. The overall mortality rate is approximately 7% (3). Although the treatment is unclear, the development of PHT may be the only indication for treatment (3). Swyer–James–Macleod syndrome, lobar atelectasis, post lobectomy status, and chronic pulmonary thromboembolism are the main differential diagnoses (6). On the other hand, pathologies that may cause deterioration of the clinical condition (acute pulmonary embolism, CTEPH, anemia, etc.) should not be ignored in such patients who have been asymptomatic for many years. When there is the presence of coronary-artery-to-pulmonary or -bronchial artery collaterals such as in our case, CTEPH should be kept in mind as a differential diagnosis. However, our case is important as, it shows that a congenital anomaly may be present in advanced age, like in the case of our patient. Kepez et al. (7) demonstrated that the rate of occurrence of coronary to PA collateral circulation was 18.1%, especially in patients with total occlusions at the level of the main pulmonary arteries (7). UAPA with coronary collaterals is a very rare anomaly, and many cases of collateral circulating are linked with RCA (8).

In a case report similar to ours with coronary-to-pulmonary collateral circulation, endothelin receptor antagonists (bosentan) for PHT were started by the authors and they mentioned this therapy is effective (9). We also showed that phosphodiesterase V inhibitors may be effective over a short period. However, considering the mechanism of development of PHT in this patient group, riociguat, which is indicated for groups 1 and 4 PHT, may be considered as the first-line treatment. One of the interesting aspects of our patient is that she is at advanced age and is similar to cases of Group 2 PHT phenotypically due to her accompanying additional comorbidities. Another interesting aspect is that her case suggests CTEPH in Group 4 PHT due to risk factors such as age, hypertension, and atrial fibrillation.

Conclusion

UAPA should be kept in mind before closing coronary-to-pulmonary-artery collaterals via surgical or percutaneous interventions.

Informed consent: Written informed consent to publication was obtained from the patient.

Video 1-2. Right coronary angiography demonstrates a normal right coronary artery with collaterals from the conus branch to the right lung tissue.

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