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CASE REPORT
Congenital heart disease

Long-term follow-up of an adult with left pulmonary artery stenting and absent right pulmonary artery: a case report

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Background Unilateral absence of pulmonary artery is a rare congenital abnormality with varied clinical presentations. We present a unique case of congenital absence of right pulmonary artery (RPA) with left pulmonary artery (LPA) origin stenosis without any intracardiac or extracardiac lesion and its long-term follow-up.

Case summary A 68-year-old woman underwent successful LPA stenting 23 years back for absent RPA and LPA origin stenosis. She was recently evaluated for breathlessness, almost two decades after the procedure. On evaluation, she was found to have severe pulmonary hypertension (Group 5) without any distortion of the LPA and with normally functioning stent.

Discussion This report discusses the long-term outcome of stenting in the setting of severe stenosis of a single pulmonary artery. Over the years, the patient went on to develop severe segmental pulmonary hypertension (Group 5).

Keywords Congenital absence of right pulmonary artery (RPA) • Breathlessness • Left pulmonary artery (LPA) stenting • Pulmonary hypertension • Case report

Learning points

- The Palmaz stent is robust and effective in the long-term treatment of pulmonary artery stenosis.
- Pulmonary hypertension can develop over years in such cases with single pulmonary artery and is segmental (Group 5) in nature.
- There is a role of pulmonary vasodilators for symptomatic relief in treatment for segmental pulmonary hypertension.

Introduction

In 1868, Frentzel[1] first reported the case of absence of right pulmonary artery (RPA). Patients with isolated unilateral absence of RPA may remain asymptomatic till late adulthood. Occasionally they can present with pulmonary hypertension and congestive heart failure in infancy.[2,3] We present an uncommon case of isolated unilateral absence of RPA with left pulmonary artery (LPA) origin stenosis which was stented and subsequently followed up for over two decades.

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Timeline

April 1997	Progressive dyspnoea for previous 3 years evaluated and underwent left pulmonary artery (LPA) stenting for LPA origin stenosis
2016–17	Reappearance of breathlessness
January 2018	Cardiac catheterization and coronary angiogram revealed high pulmonary arterial pressures without any gradient across the stent and normal coronaries
January 2018	Counselled and discharged on pulmonary vasodilators

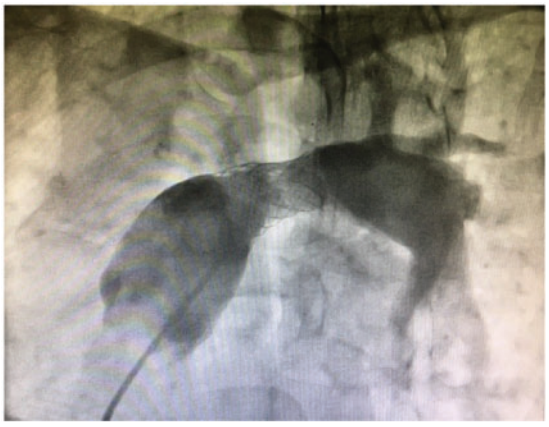

Figure 1 Pulmonary artery angiogram in left anterior oblique cranial view showing absence of the right pulmonary artery with presence of well dilated left pulmonary artery stent *in situ* with good size hilar left pulmonary artery. There is no distortion, fracture, or in-stent stenosis of the left pulmonary artery stent.

Case presentation

A 68-year-old woman presented with a 2-year history of shortness of breath and easy fatigability without any clinical evidence of right ventricular dysfunction. She underwent LPA stenting 23 years ago, for severe LPA origin stenosis with absent RPA.[4] She had a Palmaz iliac stent, mounted on an 18 mm × 4 cm balloon and passed through a 14-Fr Mullin's sheath (Cook Inc., Bloomington, IN, USA), deployed in the LPA. The internal diameter of the LPA post stenting measured 15 mm. She was discharged on Aspirin 150 mg/day for a period of 1 year.

On examination, her vitals were normal. Her cardiovascular evaluation revealed no evidence of systemic venous congestion, normal apex beat, normal first heart sound but a loud P2 without any murmur. Her lung fields were clear on auscultation. Her transthoracic echocardiography revealed suboptimal images but septae were found to be intact, valves were functioning normally with right ventricular hypertrophy, and normal ventricular systolic function (tricuspid annular plane systolic excursion 2.1 cm). There was some suggestion of turbulence across the LPA but spectral Doppler signals were non-diagnostic due to poor windows. It was, therefore, decided to perform a coronary angiogram and right heart catheter study with a view to assess her pulmonary haemodynamics.

Her cardiac catheterization revealed a right ventricular systolic pressure of 70 mmHg (systemic pressure of 130 mmHg). The LPA was crossed using a 6-Fr JR catheter, and pressures were taken across the stented LPA. There was a gradient of 5 mmHg across the stented LPA. The distal LPA pressures were 65/22 mmHg. The mean pulmonary artery pressure and mean pulmonary capillary wedge pressure were 37 mmHg and 10 mmHg, respectively. The main pulmonary angiogram done in the left anterior oblique view with a cranial angulation revealed unobstructed flow across the LPA (*Figure 1*). The stent was well positioned without any evidence of fractures or in-stent narrowing. The diameter of proximal and hilar LPA were 14 mm and 17.6 mm, respectively (*Figure 1*). Coronary angiogram revealed normal coronaries. In view of elevated PA pressures in the absence of left sided heart disease, she was diagnosed with pulmonary arterial hypertension (Group 5) and was started on pulmonary vasodilators (Sildenafil + Ambrisentan). At 3-month follow-up, the patient was symptomatically better. She is yet to undergo a repeat haemodynamic study.

Discussion

Isolated unilateral absence of a pulmonary artery (PA) is a rare entity with a prevalence of 1:200 000. Twenty-five percent of patients with unilateral absence of pulmonary artery continue to have an excellent long-term outcome due to normal or mildly elevated pulmonary artery pressures in unaffected lung.[2,5] Forty percent of the individuals with this anatomy have recurrent pulmonary infections, decreased exercise tolerance and exertional dyspnoea,[2] whereas 15% of them will remain completely asymptomatic as seen in our patient following LPA stenting. Yet another symptom, haemoptysis, which is seen in 20% of the affected individuals,[5,6] was absent in our patient. In our case, the patient did not have any large major aorto-pulmonary collateral to right lung. The pulmonary arterial hypertension in this case is most probably segmental (Group 5 pulmonary arterial hypertension).[6,7] For such cases of high PA pressures with normal pulmonary capillary wedge pressure, the use of pulmonary vasodilators will be beneficial.[6,7]

Pulmonary artery stenting was first introduced in 1991.[8] The most commonly used stent in the early era of stenting was the Palmaz iliac stent. Although it was high profiled, rigid, and non-conformable, it was robust with a high radial strength as demonstrated by our patient. There have been a large number of reports demonstrating excellent mid- and long-term results of central PA stenting.[9] Our case is one more addition to the previously reported cases with such a long follow-up. However, in view of the absence of the RPA, our case is unique.

Such patients need treatment with pulmonary vasodilators and regular follow-up to assess clinical and haemodynamic improvement.

Conclusion

This case report focusses on segmental pulmonary arterial hypertension in cases of a single pulmonary artery, and the benefits of pulmonary vasodilators in these subsets. We find that the Palmaz stent is effective in the long-term treatment of branch PA stenosis.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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