

CORONARY, PERIPHERAL, AND STRUCTURAL INTERVENTIONS

CLINICAL CASE

Long-Term Sequential Percutaneous Treatment in Takayasu's Arteritis With Pulmonary Artery Involvement as the Initial Presentation



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ABSTRACT

BACKGROUND Takayasu's arteritis (TA) is a rare inflammatory disease affecting large vessels, with pulmonary artery involvement being uncommon and challenging to manage.

CASE SUMMARY A 46-year-old woman with severe pulmonary hypertension, initially diagnosed with TA, presented with left pulmonary artery stenosis and right pulmonary artery total occlusion. After a series of percutaneous interventions over 20 years, the patient avoided lung transplantation and achieved stable long-term outcomes.

DISCUSSION This case is of particular interest due to the rare pulmonary involvement in TA and the successful management of chronic total pulmonary artery occlusion through sequential percutaneous interventions. The extended 20-year follow-up is exceptional, as long-term outcomes in similar cases are rarely documented.

TAKE-HOME MESSAGE Percutaneous treatments can provide significant long-term benefits in managing pulmonary artery involvement in TA, even in complex cases. (JACC Case Rep. 2025;30:104132) © 2025 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

HISTORY OF PRESENTATION

A 46-year-old woman presented 20 years ago with dyspnea and chest pain on minimal exertion, in NYHA functional class III-IV. Pulmonary arterial hypertension (PAH) was diagnosed, and the patient was referred to our center for lung transplantation. At the time of

TAKE-HOME MESSAGE

- Percutaneous treatment can offer sustained long-term benefits for pulmonary artery stenosis in patients with pulmonary hypertension due to Takayasu's arteritis, even in complex cases.

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**ABBREVIATIONS
AND ACRONYMS**

LPA = left pulmonary artery
PA = pulmonary artery
PAH = pulmonary arterial hypertension
PFO = patent foramen ovale
RA = right atrium
RHC = right heart catheterization
RPA = right pulmonary artery
RV = right ventricle
TA = Takayasu's arteritis
TTE = transthoracic echocardiography

presentation at our center, physical examination revealed central cyanosis and systolic murmurs over the carotid and subclavian arteries, as well as in the pulmonic area.

PAST MEDICAL HISTORY

Previous medical history was unremarkable.

DIFFERENTIAL DIAGNOSIS

Based on the clinical presentation, differential diagnoses could include idiopathic PAH, Eisenmenger syndrome, and chronic thromboembolic pulmonary hypertension. The presence of vascular bruits raised suspicion for large-vessel vasculitis, particularly Takayasu's arteritis (TA). Other considerations included left-sided heart disease and rare systemic vascular disorders affecting the pulmonary circulation.

INVESTIGATIONS

A chest radiograph showed marked oligemia in both lungs. Electrocardiography indicated right axis deviation with right ventricular enlargement. Transthoracic echocardiography (TTE) identified right ventricular dilation with mild dysfunction and an estimated systolic pulmonary artery (PA) pressure of 130 mm Hg, with no signs of valvular disease and a normal left ventricular ejection fraction. The results of pulmonary function tests were normal, and the 6-minute walking test distance was 300 m, with marked oxygen desaturation, dropping from 90% at rest to 76% during exercise, attributed to a patent foramen ovale (PFO). A study of the PFO was performed, which showed a moderate right-to-left shunt after bubble

TABLE 1 Timeline of Clinical Status and Percutaneous Treatments Along 20 Years

Timeline	Events
26 y	Dyspnea, NYHA functional class III-IV RHC with PH and PA stenosis. Diagnosis of TA Stent in LPA Steroids and azathioprine
41 y	Recurrence of dyspnea. NYHA functional class III RHC. Chronic occlusion of RPA stenting. Postdilatation stent LPA Tocilizumab
46 y	Recurrence of dyspnea. NYHA functional class III RHC. Restenosis in LPA and mismatch in RPA. Postdilatation Steroids

LPA = left pulmonary artery; PA = pulmonary artery; PH = pulmonary hypertension; RHC = right heart catheterization; RPA = right pulmonary artery; TA = Takayasu's arteritis.

TABLE 2 Timeline of Hemodynamic and Echocardiographic Data

Timeline	Details
26 y	
RHC	
Basal	LPAP 30/10/16, PAP 105/18/46, LPAPg 75, RVP 102/14/20, RAP 12, LAP 5, CI 1.9, $Q_p/Q_s = 0.86$
Post	LPAP 41/18/25, PAP 56/22/33, LPAPg 15, RVP 55/9/11, RAP 7, LAP 5, CI 2.3, $Q_p/Q_s = 1$
Echo	
Basal ^a	TAPSE 14, RV-RAG 110, TR grade II, RVsize 46 3 mm PFO without ASA, moderate shunt with bubbles
Post ^b	TAPSE 23, RV-RAG 30, TR grade I, RV size 37. Mild right-to-left shunt after Valsalva with bubbles
41 y	
RHC	
Basal	LPAP 38/9/21, PAP 132/14/53, LPAPg 95, RVP 132/4/15, RAP 10, LAP 4, CI 1.7
Post	RPAP 46/14/25, LPAP 53/13/26, PAP 64/15, RPAPg 18, LPAPg 11, RVP 63/6/11, RAP 7, LAP 3, CI 2.4
Echo	
Basal ^a	TAPSE 13, RV-RAG 122, TR grade II, RV size 52
Post ^b	TAPSE 20, RV-RAG 30, TR grad I, RV size 37
46 y	
RHC	
Basal	RPAP 54/14, LPAP 41/16/24, PAP 67/12/26, LPAPg 26, RPAPg 13, RVP 70/9/13, RAP 10, LAP 3
Post	LPAP 39/10/24, RPAP 48/13/26, PAP 58/12, LPAPg 19, RPAPg 10, RVP 58/6/10, RAP 7, LAP 4
Echo	
Basal ^a	TAPSE 17, RV-RAG 70, TR grade I, FAC-RV 36%, RV size 46
Post ^b	TAPSE 22, RV-RAG 49, TR grade I, FAC-RV 41%, RV size 41

^aPrevious to catheterization. ^bFirst 6 months after catheterization.

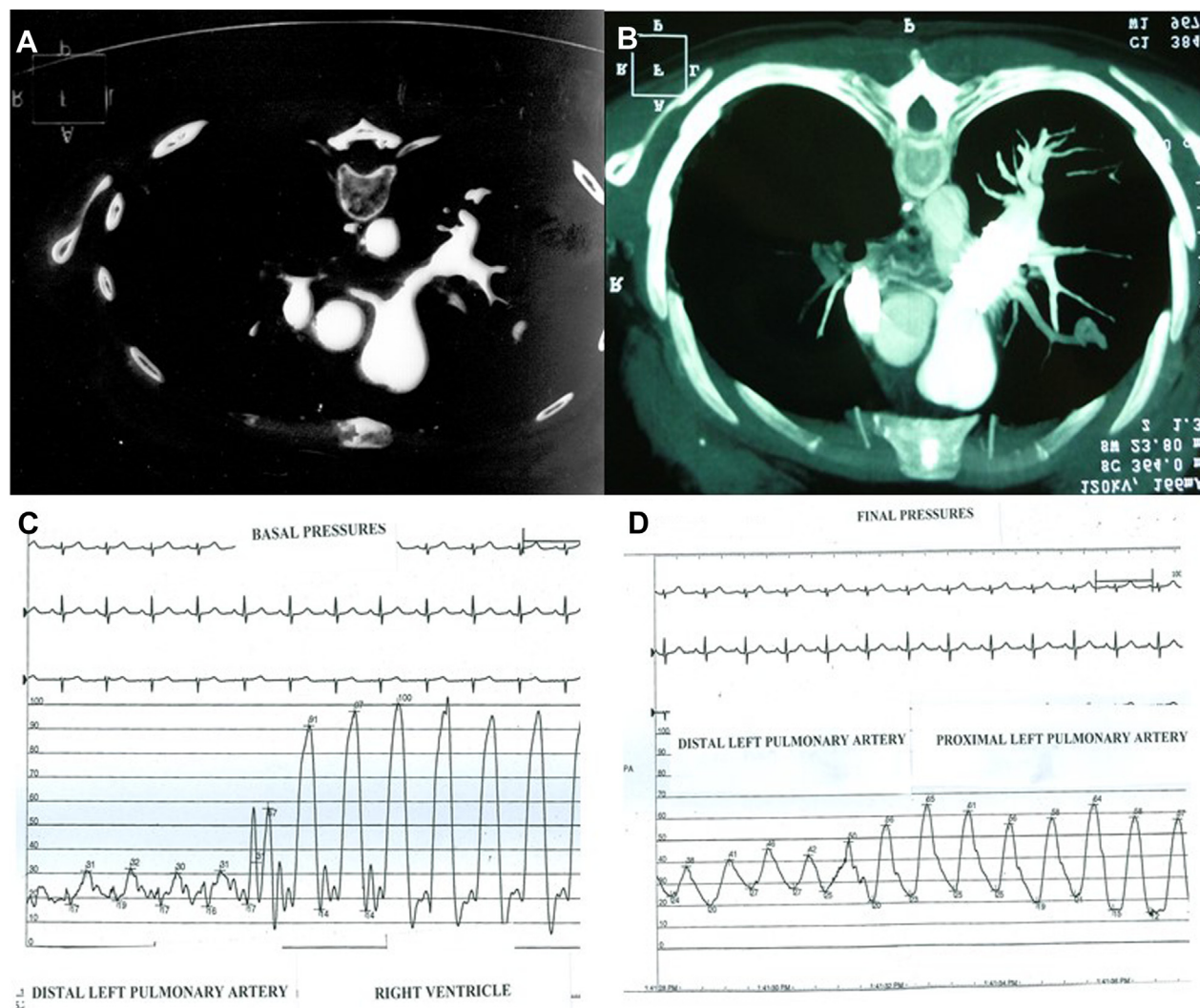
ASA = aneurysm of the interatrial septum; CI = cardiac index (L/min/m² body surface area, by the indirect Fick method); FAC-RV = fractional area change of the right ventricle (%); LAP = left atrial pressure (mm Hg, obtained through the PFO); LPAPg = left pulmonary artery gradient (mm Hg, peak-to-peak); LPAP = left pulmonary artery pressure (mm Hg, distal to the obstruction); PAP = pulmonary artery pressure (mm Hg, pulmonary trunk); PFO = patent foramen ovale; $Q_p/Q_s = 1$ (pulmonary flow/systemic flow); RAP = right atrial pressure (mm Hg); RHC = right heart catheterization; RPAPg = right pulmonary artery gradient (mm Hg, distal to the obstruction); RV-RAG = right ventricle-right atrium gradient (mm Hg); RVP = right ventricular pressure (mm Hg); RV size = size of the right ventricle (basal diameter in mm, 4-chamber RV-focused view); TAPSE = tricuspid annular plane systolic excursion (mm); TR = tricuspid regurgitation (I-IV).

administration, with an opening size of 3 mm and no aneurysm of the interatrial septum. Lung ventilation and perfusion scan showed markedly reduced perfusion in the left lung, absent perfusion in the right lung and normal ventilatory patterns. A magnetic resonance angiography revealed an increased thickness of the aortic thoracic wall, stenosis of the right subclavian artery, stenosis of the left main PA (LPA), and total occlusion of the right PA (RPA). TA with pulmonary involvement was confirmed.

MANAGEMENT

The timeline for the sequential pulmonary arterial angioplasties is shown in [Table 1](#). [Table 2](#) presents the

FIGURE 1 Computed Tomography and Hemodynamic Evaluation Before and After Stent Placement in the Left Pulmonary Artery

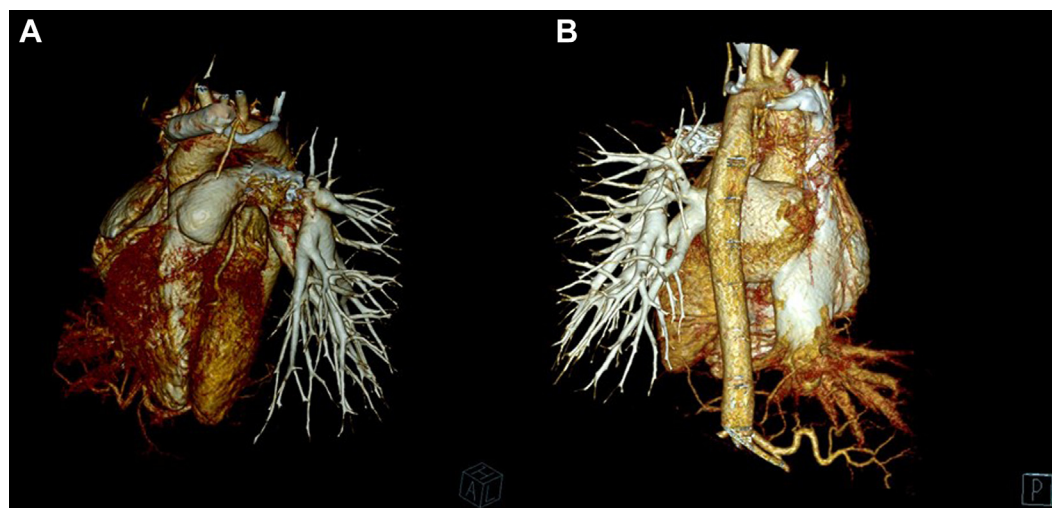


(A) Contrast-enhanced axial CT scan depicting the severe left pulmonary artery stenosis and the occlusion of the right pulmonary artery. (B) Axial CT depicting the deployed stent in the left pulmonary artery. (C) Basal hemodynamic pullback from the left pulmonary artery to the right ventricle depicting the severe gradient between the right ventricle and distal left pulmonary artery. (D) Final hemodynamic pullback from the distal to proximal left pulmonary artery showing only a mild gradient across the stent. CT = computed tomography.

pre- and postangioplasty hemodynamic values, along with the echocardiographic parameters.

Initial right heart catheterization (RHC) confirmed a low systemic cardiac index (1.9 L/min/m² body surface area) with a Q_p/Q_s ratio of 0.86, a main PA pressure of 105/18 mm Hg (mean 46 mm Hg), and a distal LPA pressure of 30/10 mm Hg (peak-to-peak systolic pressure gradient 75 mm Hg), indicating severe stenosis of the LPA. The RPA was totally occluded (Figure 1).

The LPA stenosis was predilated, and a 34-mm 8 zig CP Stent (NuMED Inc) was then successfully deployed and postdilated up to 18 mm diameter across the stenosis. The peak-to-peak systolic pressure gradient across the stenosis decreased markedly to 15 mm Hg, with no clinical or angiographic complications (Figures 1 and 2). Normalization of oxygen saturation was observed, with a Q_p/Q_s ratio of 1 and a mild right-to-left shunt after the Valsalva maneuver with the bubble test.

FIGURE 2 3D Computed Tomography Reconstruction of the Pulmonary Arterial Anatomy After Left Pulmonary Artery Stent Placement

3D reconstruction of the pulmonary arterial tree from a computed tomography angiogram (A: anterior view, B: posterior view) showing the absence of vascularization in the right lung.

OUTCOMES AND FOLLOW-UP

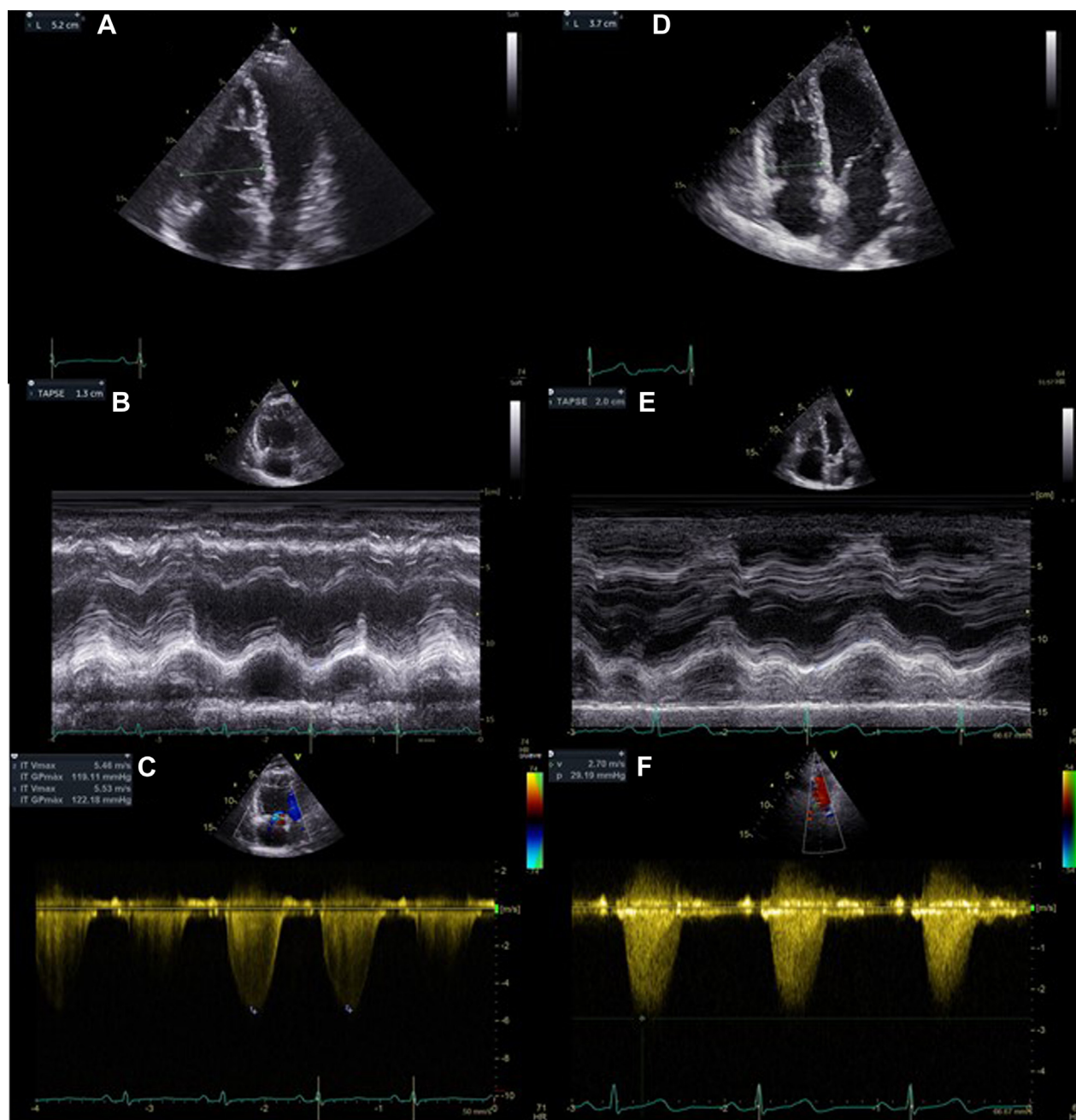
Over a 10-year follow-up period, the patient remained stable in NYHA functional class I-II, with a 6-minute walking test distance of 480 m without desaturation and a systolic PA pressure of 35 mm Hg on TTE.

RHC was performed 15 years after the initial procedure due to increased exertional dyspnea, worsening right ventricular function (tricuspid annular plane systolic excursion 13 mm, ventricular dilation with a basal diameter of 52 mm), and an increase in the right ventricle-right atrium (RV-RA) gradient (Figures 3A to 3C show the echocardiographic findings). An angiographic and hemodynamic assessment revealed that restenosis of the previously implanted stent in the LPA and persistent total occlusion of the RPA were observed. Stent restenosis was postdilated with a 16 × 20 mm high-pressure Atlas Gold balloon (Bard Peripheral Vascular, Inc). Subsequently, the occlusion of the RPA was treated using the step-by-step process illustrated in Figure 4. The occluded RPA was successfully crossed using coronary guidewires designed for total occlusion (MIRACLEBros 6, Asahi Intecc Co, Ltd), reaching the distal right lower lobe pulmonary artery. Distal patency was confirmed by visualizing the distal pulmonary tree beyond the occlusion site, with no signs of thrombotic material or new obstructive lesions. A 7 × 20 mm Conquest balloon (Bard Peripheral Vascular, Inc) was used for

predilatation. Two overlapping stents were deployed: a PALMAZ Blue 6 × 18 mm PALMAZ Blue 6 × 18 mm bare-metal stent (Cordis, Cardinal Health) and an Advanta 6 × 38 mm covered stent (Atrium Medical Corporation, Getinge Group), followed by post-dilatation with an 8 × 20 mm Conquest balloon (Bard Peripheral Vascular, Inc) (Figure 4). The final PA pressure was 46/14 mm Hg (mean 25 mm Hg) and the left atrial pressure through the PFO 3 mm Hg. The procedure was uneventful and the patient showed marked improvement, achieving NYHA functional class I with normalization of right ventricular function, dilation, and the RV-RA gradient (Figures 3D to 3F). The ventilation-perfusion parameters showed reperfusion of the right lower and middle lobes and improvement in left-sided perfusion (Figure 5).

At the 20-year follow-up, the patient returned to NYHA functional class III, and an increased RV-RA gradient of 70 mm Hg was observed on TTE. Positron emission tomography scan showed inflammatory activity in the LPA stent (maximum standardized uptake value 7.6), without any activity in the RPA stent (18F-fluorodeoxyglucose) (Figure 6). At the same time, an increase in inflammatory markers, which had previously been normal, was observed, with a peak of C-reactive protein at 8.3 mg/dL (normal value 0.5 mg/dL), an erythrocyte sedimentation rate of 108 mm/h (normal value <20 mm/h), and an interleukin-6 level of 31.2 pg/mL (normal value

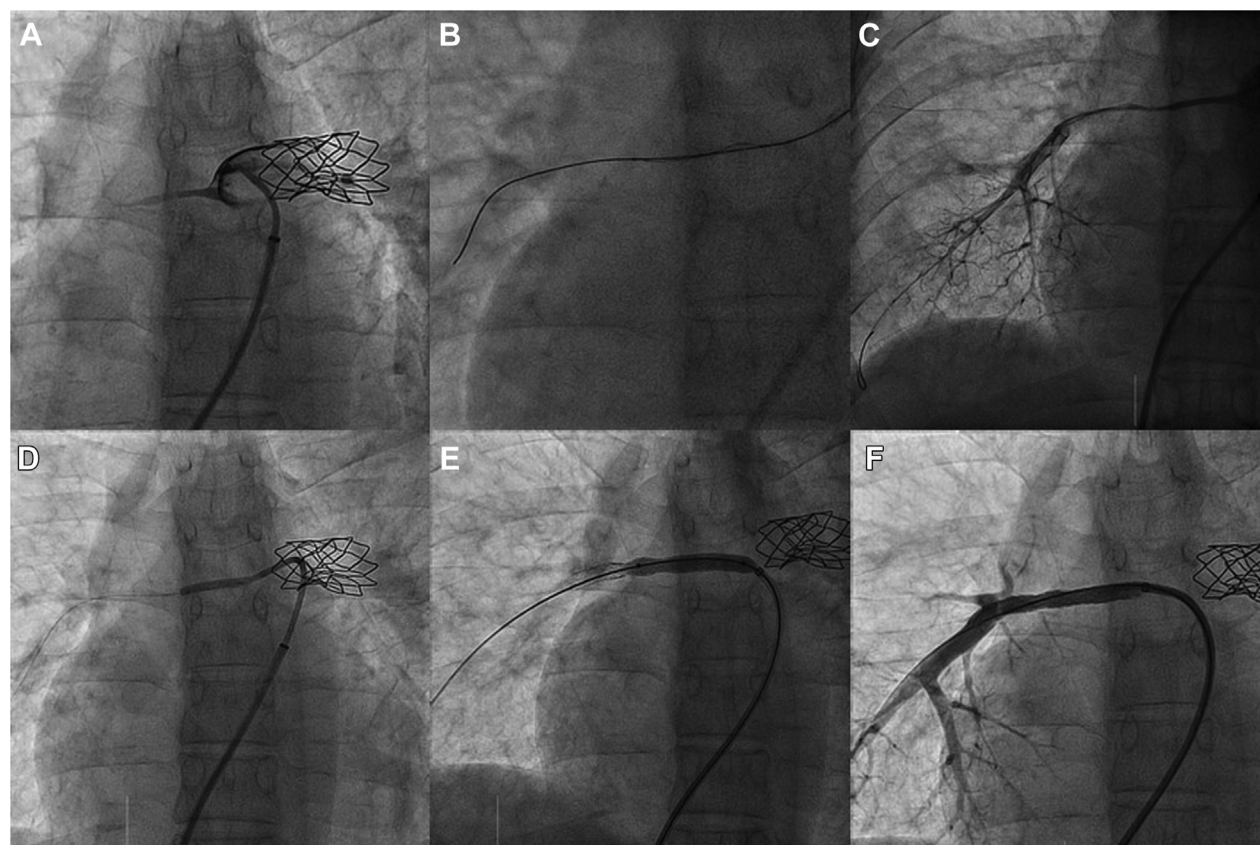
FIGURE 3 Echocardiographic Evolution Before and After Treatment of Right Pulmonary Artery Occlusion



Echocardiographic findings before (A-C) and after (D-F) the second procedure (postdilation of the LPA and opening of the RPA occlusion). (A) Dilated right ventricle with the interventricular septum shifted to the left and a basal diameter of 52 mm. (B) TAPSE of 13 mm. (C) RV-RA gradient of 122 mm Hg. (D) Normalization of right ventricular dilation with a centered septum (basal diameter of 37 mm). (E) TAPSE of 20 mm. (F) RV-RA gradient of 29 mm Hg. LPA = left pulmonary artery; RA = right atrium; RPA = right pulmonary artery; RV = right ventricle; TAPSE = tricuspid annular plane systolic excursion.

<4.3 pg/mL). A new RHC showed a peak-to-peak gradient of 26 mm Hg across the LPA and 13 mm Hg through the RPA stent. Angiographic restenosis, likely inflammatory, was seen in the LPA, along with a

reduced diameter due to mismatch in the RPA stent. The RPA and LPA stents were postdilated with 12 × 20 mm and 14 × 20 mm Atlas Gold balloons (Bard Peripheral Vascular, Inc), respectively (Figure 7).

FIGURE 4 Step-by-Step Procedure for the Treatment of Right Pulmonary Artery Occlusion

(A) Baseline angiography shows the occlusion. (B) Guidewire passage through the occlusion. (C) Distal angiography visualizing the vasculature distal to the lesion. (D) Predilation. (E) Deployment of 2 overlapping stents. (F) Final angiography confirming vessel patency.

Final hemodynamic pressures in mm Hg were as follows: aortic 107/52, RV 58/6, distal RPA 48/13 (mean 26), distal LPA 39/10 (mean 24), and LA 4. Intravascular ultrasound of the distal pulmonary artery was performed to assess wall thickening, revealing a wall thickness area ratio of 25% in the distal RPA and 30% in the distal LPA (Figure 8). The patient returned to NYHA functional class II. The newly detected mild distal PA hypertension was not initially treated.

Over the years she has been sequentially treated with corticosteroids, azathioprine, and tocilizumab. Pulmonary vasodilator therapy was considered, but given the limited evidence of benefit and the patient's clinical improvement, it was not initiated.

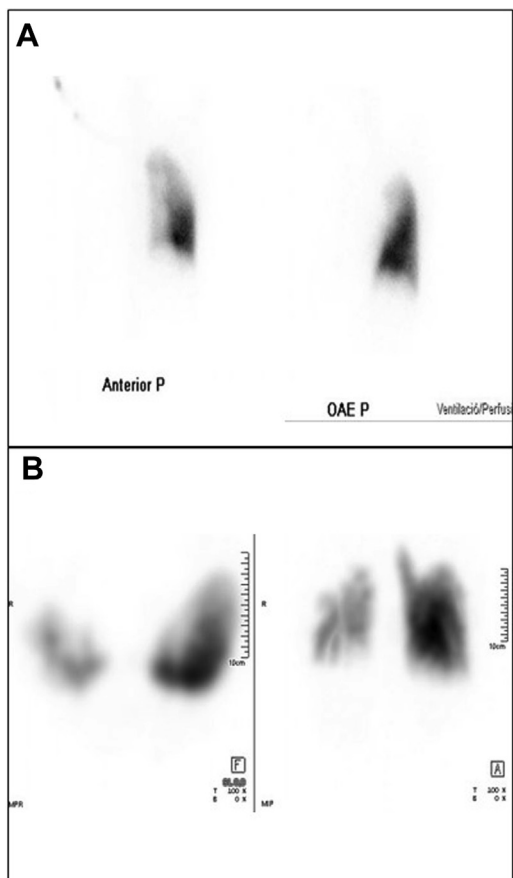
DISCUSSION

This case highlights the atypical initial presentation of TA with pulmonary involvement as the first clinical manifestation.¹ Despite this uncommon presentation,

the application of the American College of Rheumatology/EULAR (2022) classification criteria yields a score exceeding 5, supporting the diagnosis of TA.² The patient meets several key criteria, including young age, female sex, presence of a carotid bruit, carotid artery stenosis, and involvement of 3 distinct arterial territories (aorta, pulmonary, and carotid arteries). Furthermore, evidence of vasculitis on positron emission tomography-computed tomography imaging and elevated acute phase reactants provides additional diagnostic support. Up to a quarter of patients with TA exhibit pulmonary involvement, with stenosis being the most common presentation and only 16% developing severe pulmonary hypertension.¹

A mortality rate of up to 20% has been reported in patients with pulmonary arteritis and 33% in those with PAH.^{3,4} Although first-line treatment remains medical therapy⁵ with glucocorticoids combined with immunosuppressants or biologics, percutaneous treatment may be considered for some patients with

FIGURE 5 Ventilation-Perfusion Scan Before and After Revascularization



(A) Ventilation perfusion scan showing the absence of perfusion in the right lung. (B) Ventilation perfusion scan showing marked improvement of perfusion in the right lung.

severe or bilateral involvement.^{3,4,6} The most feared short-term complication of pulmonary angioplasty is reperfusion pulmonary edema, occurring in up to half of the patients.⁶ For this reason, the RPA was not initially treated.

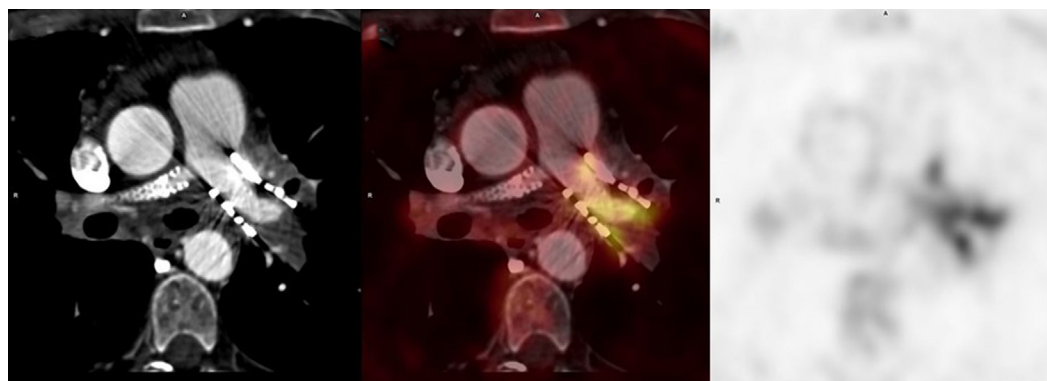
The restenosis rate seems lower when vascular interventions are performed at the stable stage of the disease, rather than during the active inflammatory phase. In addition, the use of stent implantation has significantly reduced these rates compared with simple balloon angioplasty.³ However, very long-term follow-up has not been widely reported, and it is reasonable that TA, as a chronic disease, may require several sequential interventions throughout a patient's lifetime.

In addition, clinical and angiographic successful percutaneous treatment of a PA chronic total occlusion in this context has not been previously reported.⁷

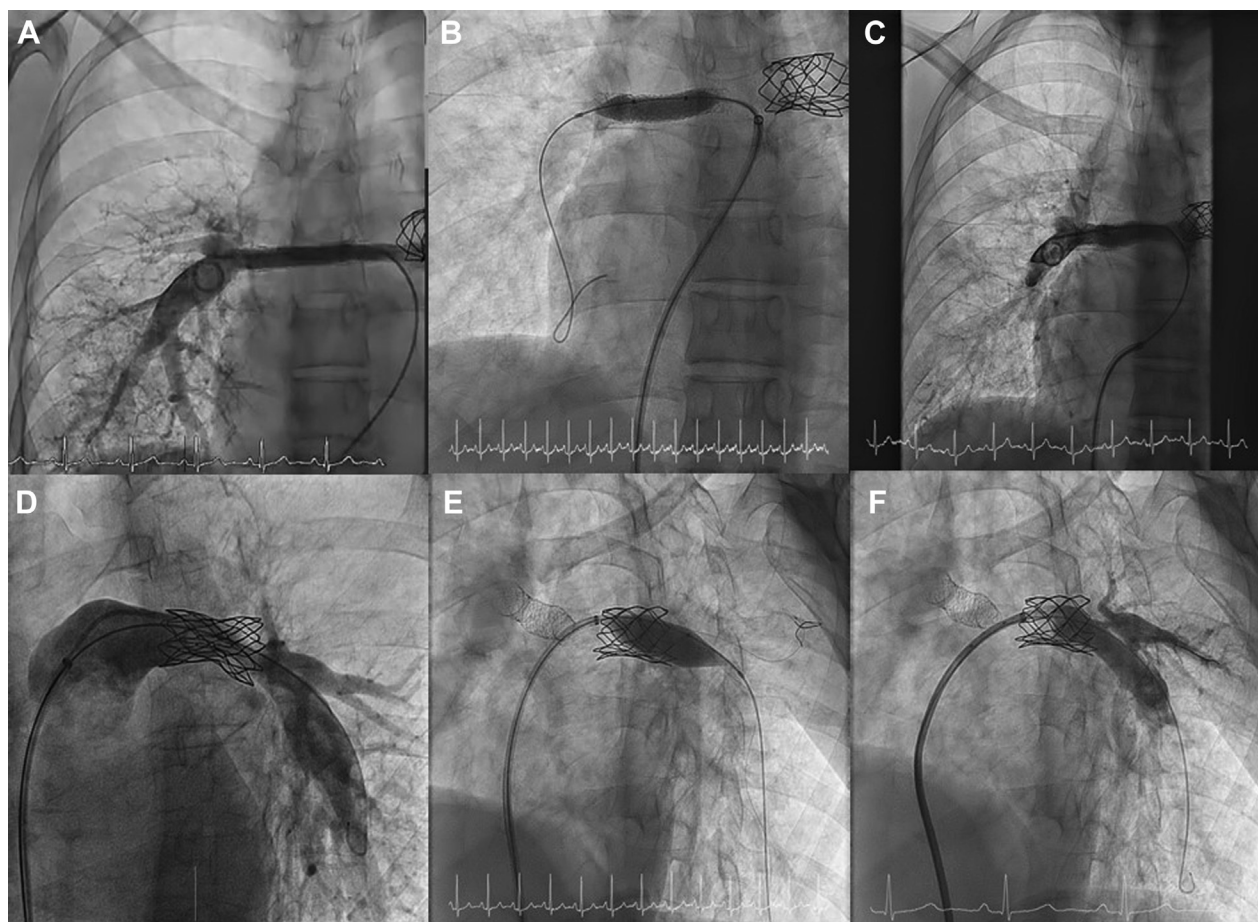
The clinical improvement can be attributed to better ventilation-perfusion matching and a decreased right ventricular afterload improving cardiac output. Although the PFO may provide hemodynamic benefits by relieving right atrial pressure, it can also worsen cyanosis. This cyanosis resolves after percutaneous treatment of the stenosis, improving oxygenation due to a reduction in right-to-left shunting.⁸

The development of distal pulmonary hypertension at follow-up is not usual and may be due to inflammatory small vessel disease or segmental stenosis of distal vessels, leading to reactive pulmonary hypertension, suggesting a potential role for pulmonary vasodilators. Although some benefit has been reported in selected cases, its efficacy remains uncertain.^{5,9}

FIGURE 6 6-Fluorodeoxyglucose Positron Emission Tomography Scan Images of the Pulmonary Arteries



Positron emission tomography scan showing inflammatory activity in the left pulmonary artery branch stent with 6-fluorodeoxyglucose uptake (maximum standardized uptake value of 7.6) and no uptake in the right pulmonary artery branch stent. A mismatch in right branch size is observed, with a smaller diameter in the right pulmonary artery.

FIGURE 7 Pulmonary Artery Fluoroscopic Angiographies From the Last Procedures

Contrast angiography of pulmonary arteries. (A) Mismatch stenosis of the right pulmonary artery. (B) Balloon dilatation of the right pulmonary artery. (C) Final result in the right pulmonary artery. (D) Severe intrastent restenosis in the left pulmonary artery. (E) Balloon dilatation in the left pulmonary artery. (F) Final result in the left pulmonary artery.

CONCLUSIONS

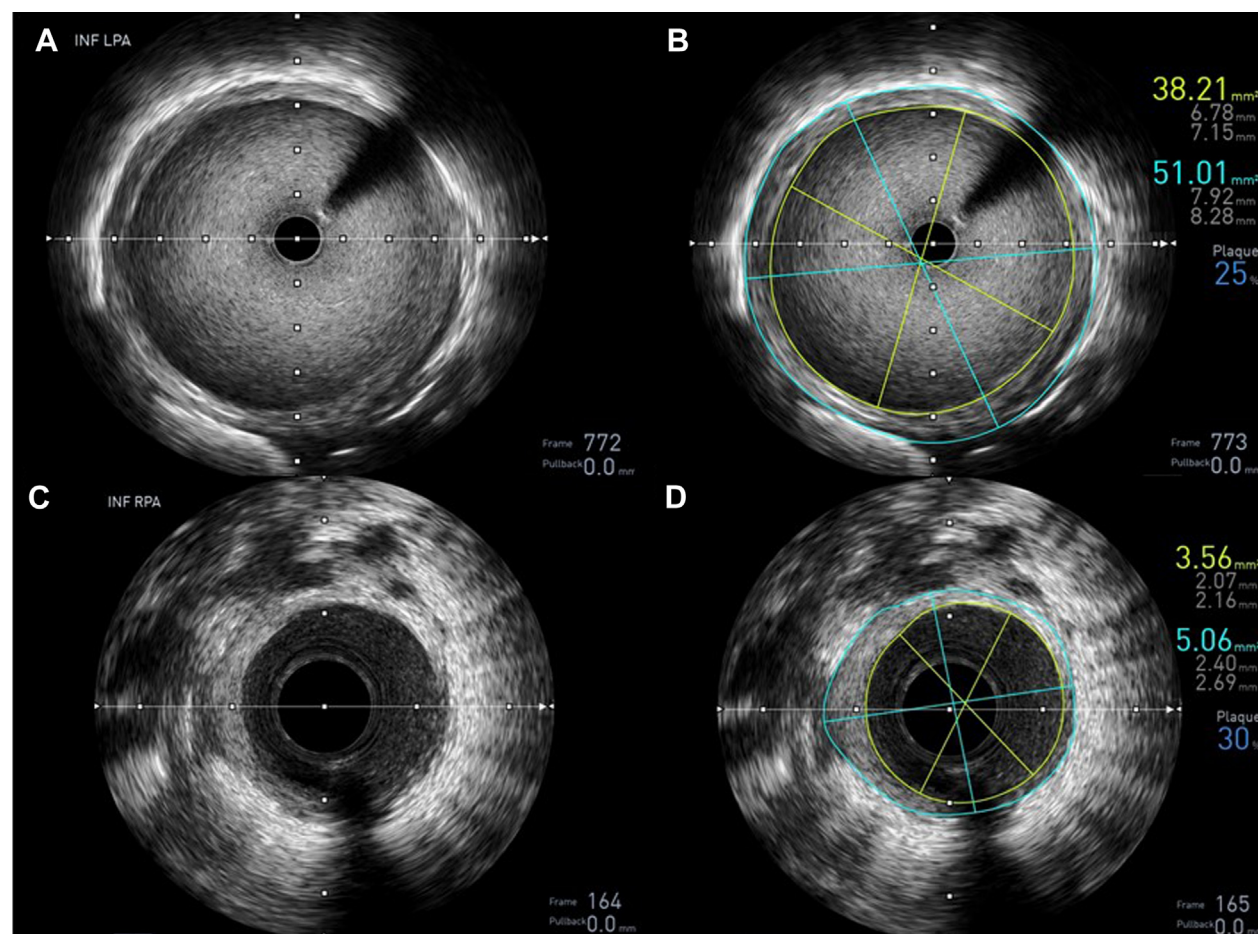
The optimal indications and timing for percutaneous treatment of pulmonary artery stenosis in Takayasu's disease remain unclear. However, this approach seems to be a viable option for selected patients, even with chronic total occlusions, showing favorable long-term outcomes.

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FIGURE 8 Intravascular Ultrasound Imaging in Distal Pulmonary Arteries



Intravascular ultrasound imaging in the distal pulmonary arterial tree showing remodeling of the pulmonary arterial wall (thickening). Remodeling in the inferior left pulmonary artery (A, B) and remodeling in the inferior right pulmonary artery (C, D).

REFERENCES

- Kong X, Ma L, Lv P, et al. Involvement of the pulmonary arteries in patients with Takayasu arteritis: a prospective study from a single centre in China. *Arthritis Res Ther*. 2020;22(1):131.
- Grayson PC, Ponte C, Suppiah R, et al. 2022 American College of Rheumatology/EULAR classification criteria for Takayasu arteritis. *Ann Rheum Dis*. 2022;81(12):1654-1660.
- Zhou YP, Wei YP, Yang YJ. Percutaneous pulmonary angioplasty for patients with Takayasu arteritis and pulmonary hypertension. *J Am Coll Cardiol*. 2022;79(15):1477-1488.
- Qin L, Hong-Liang Z, Zhi-Hong L, Chang-Ming X, Xin-Hai N. Percutaneous transluminal angioplasty and stenting for pulmonary stenosis due to Takayasu's arteritis: clinical outcome and four year follow-up. *Clin Cardiol*. 2009;32:639-643.
- Fukamoto Y. Takayasu arteritis-associated pulmonary hypertension. *Eur Heart J*. 2021;42(42):4306-4308.
- Inami T, Kataoka M, Shimura N, et al. Pulmonary edema predictive scoring index (PEPSI), a new index to predict risk of reperfusion pulmonary edema and improvement of hemodynamics in percutaneous transluminal pulmonary angioplasty. *JACC Cardiovasc Interv*. 2013;6(7):725-736.
- Hong-Mi C, Hyung-Kwan K, Hyo-Sun S, et al. Total occlusion of right main pulmonary artery in a patient with Takayasu arteritis and severe pulmonary hypertension. *J Cardiovasc Ultrasound*. 2012;4:189-192.
- Sharan L, Stackhouse K, Awerbach JD, Bashore TM, Krasuski RA. Effect of patent foramen ovale in patients with pulmonary hypertension. *Am J Cardiol*. 2018;122(3):505-510.
- Nitzberg M, Parikh R, Govender P, Farber HW. Pulmonary hypertension secondary to Takayasu's arteritis: management using a combined medical and interventional approach. *Sarcoidosis Vasc Diffuse Lung Dis*. 2020;37(2):239-241.

KEY WORDS cardiac catheterization, pulmonary artery stenosis, pulmonary hypertension, pulmonary stenting, Takayasu