Comparison of long-term survival after endovascular treatment *versus* medical therapy in patients with Takayasu's arteritis and pulmonary artery stenosis

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Abstract

Objective

There is currently no unanimous consensus on the treatment of Takayasu's arteritis (TAK) involving the pulmonary artery, and there are very few related studies that compare the efficacies of drug-based therapy and revascularisation. This study aimed to compare the long-term survival outcomes after endovascular treatment and medical therapy in TAK patients with pulmonary artery stenosis (PAS) and pulmonary hypertension (PH).

Methods

A total of 129 TAK patients with PAS and PH (women, 101; men, 28; mean age, 40.5 years) were retrospectively enrolled in this study. Data on clinical features, treatment regimens, and mortality, were collected. Patients were categorised into medical treatment (n=75) and percutaneous transluminal pulmonary angioplasty (PTPA; n=54: 52 with PTPA and 2 with stent implantation) groups. The primary endpoint was cardiac mortality. The median follow-up time was 54 (40.5, 58.5) months.

Results

There were no significant differences in sex, age, comorbidities, disease activity, World Health Organisation (WHO) function classification, and 6-min walk distance (6MWD) between the two groups at baseline. Compared with the conservative treatment group, cardiac mortality, WHO functional class, and 6MWD were significantly improved in the PTPA group ($p=0.031$, $p<0.001$, and $p=0.004$, respectively).

Conclusion

Under basic medicine, PTPA therapy improves the long-term survival of TAK patients with PAS and PH compared to medical treatment alone. PTPA may be a promising modality for the TAK patients with PAS and PH.

Key words

Takayasu's arteritis, pulmonary artery stenosis, pulmonary hypertension, p ercutaneous transluminal pulmonary angioplasty

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Introduction

Takayasu's arteritis (TAK) is a chronic vasculitis of unknown aetiology that mainly affects the aorta and its branches (1, 2). TAK is more common in Eastern than in Western countries, and usually affects women of childbearing age. The early clinical diagnosis of TAK is extremely difficult because of the lack of specific symptoms and signs (3). The treatment of TAK is challenging, although glucocorticoid, immunosuppressive agents, and biological disease-modifying anti-rheumatic drugs could be use (4). TAK involving the pulmonary artery, causing pulmonary artery stenosis (PAS) or occlusion, is common. According to previous reports (5-8), the prevalence of pulmonary arterial involvement in patients with TAK ranges from 13.3% to 61.7% in different populations. Once the pulmonary artery is involved, it often causes pulmonary hypertension, which poses a major challenge in the treatment of TAK, resulting in a poor prognosis. Jiang et al. conducted a national registration study and found that the overall 1-, 3-, and 5-year survival rates in TAK-PH were 94.0%, 83.2%, and 77.2%, respectively (9). In the past, there was a lack of effective treatments for TAK-related pulmonary hypertension (PH), except for traditional drugs and PH-targeted drug therapy (9-11). Despite small sample studies and case reports (12-14), percutaneous transluminal pulmonary angioplasty (PTPA) for pulmonary artery stenosis in TAK patients has not been widely used in the clinical practice for a long time, due to the high surgery-associated complication rate and mortality. However, with recent advances in PTPA for chronic thromboembolic pulmonary hypertension (15), especially the reduction of acute pulmonary oedema, the safety of PTPA treatment for PAS caused by TAK has improved significantly, and the use of PTPA in the treatment of pulmonary artery stenosis associated with TAK is also increasing. In addition, some data in this study had been reported in our previous study. It indicated that PTPA may be safe and effective for the treatment of pulmonary arterial lesions induced by TAK (16). Long-term follow-up outcomes of PTPA in patients with TAK-related PAS are lacking, and very few studies have evaluated whether PTPA treatment is superior to medical treatment in such patients. Interesting, a recent study showed that PTPA tended to be associated with a reduced risk of all-cause mortality with acceptable safety profiles compared to the non-PTPA group (17). Hence, this study aimed to compare the long-term outcomes following PTPA treatment versus medical therapy in patients with Takayasu's arteritis and PAS.

Patients and methods

Design and setting

A total of 129 consecutive patients with TAK and PAS from two medical school-affiliated hospitals were retrospectively enrolled in this study between January 2016 and December 2021. The research data were obtained through the hospitals' electronic medical record system. The study was approved by the ethics committee of Fuwai Hospital and by the local IRB (no.: 2021-1520). All included patients provided informed consent.

Selection of patients

Patients with TAK who underwent contrast-enhanced computed tomography of the pulmonary arteries (CTPA) were screened. All patients with TAK fulfilled the 1990 American College of Rheumatology (ACR) criteria for TAK and most of subjects (124, 97.5%) met the 2022 ACR /European Alliance of Associations for Rheumatology (EULAR) classification criteria (18, 19). The degree of stenosis (%) was obtained by calculating the difference between the reference vessel diameter and the stenotic vessel diameter and dividing it by the reference vessel diameter. If the ratio was greater than 50%, the case was defined as PAS. Patients with TAK and PAS confirmed by CTPA, and pulmonary hypertension (PH) assessed by transthoracic echocardiography (TTE), were included, whereas patients with left heart failure were excluded from this study. PH was defined as an estimated pulmonary artery systolic pressure (PASP) of >50 mmHg and a TRV of >3.4 m/s, according to the European Society of Cardiology/European Respiratory Society guidelines (20). All selected patients

received basic drug therapy, including prednisone, immunosuppressants, or PH-targeted therapy. Remission was defined as stable disease, with normal levels of C-reactive protein (CRP) and erythrocyte sedimentation rate, and without disease activity on imaging. The therapeutic regimen with PTPA or not was jointly decided by the treating physician and patients' preference. The enrolled patients were divided into the endovascular treatment and medical therapy groups, based on whether they received PTPA treatment or not.

Data collection

The patients' medical information, including sex, age, symptoms, signs, laboratory test results, comorbidities, medication, echocardiography, findings of CTPA, interventional treatment status, and cause of death, was collected and recorded through the electronic medical system. The primary endpoint of this study was cardiac death. The outcomes of CTPA were analysed by a radiologist who was blinded to the patients' grouping. All patient data were manually checked by two of our colleagues to ensure completeness and accuracy.

Treatment and follow-up

All patients received conventional drug therapy, such as diuretics and digoxin. If needed, glucocorticoids and/or immunosuppressants were also prescribed. If the diagnosis of PH was confirmed by right heart catheterisation (RHC), PH-targeted treatment was prescribed, including phosphodiesterase (PDE) 5 inhibitors, endothelin receptor antagonist, and prostacyclin analogue. If patients refused to undergo RCH but the diagnosis of PH was supported by TTE, PH-targeted therapy was also considered. Patients who underwent PTPA needed to be in remission and without disease activity before the procedure. Those patients who underwent PTPA received aspirin (100 mg/day) and clopidogrel (75 mg/day) for at least five days before the intervention. After intervention, they would be administered dual antiplatelet therapy for three months unless severe bleeding complications. PTPA was performed via femoral vein access with local anaesthesia. An 8 F introducer sheath was inserted using the Seldinger technique. An 8 F guide catheter (MPA1or JR4) was then inserted through the sheath, and a 0.014-inch guidewire was passed through the target lesion. Unfractionated heparin was administered at a dose of 1 mg/kg to maintain the appropriate activated clotting time. Patients underwent RHC before and after PTPA and at follow-up examinations. Pulmonary artery angiography was performed before each PTCA to select and identify the target lesions but not after the intervention. Balloon size was determined by measuring vessel diameter using an imaging ruler and, occasionally, optical coherence tomography. Based on the diameter of the normal vascular segment, which included the targeted lesions, a relatively small balloon was used for balloon expansion, and then the balloon diameter was sequentially increased to obtain a larger vascular lumen and greater pulmonary artery blood flow. Patients were followed-up at 3, 6, and 12 months, and every 12 months after that. During the visits, patients were interviewed, and routine blood tests (blood routine, liver function, serum creatinine, CRP, and erythrocyte sedimentation rate), electrocardiogram, TTE, World Health Organisation (WHO) functional class, 6-min walk distance (6MWD), and medication regimen, were recorded. Patients who received PTPA treatment were suggested to undergo RHC 12 months after the intervention.

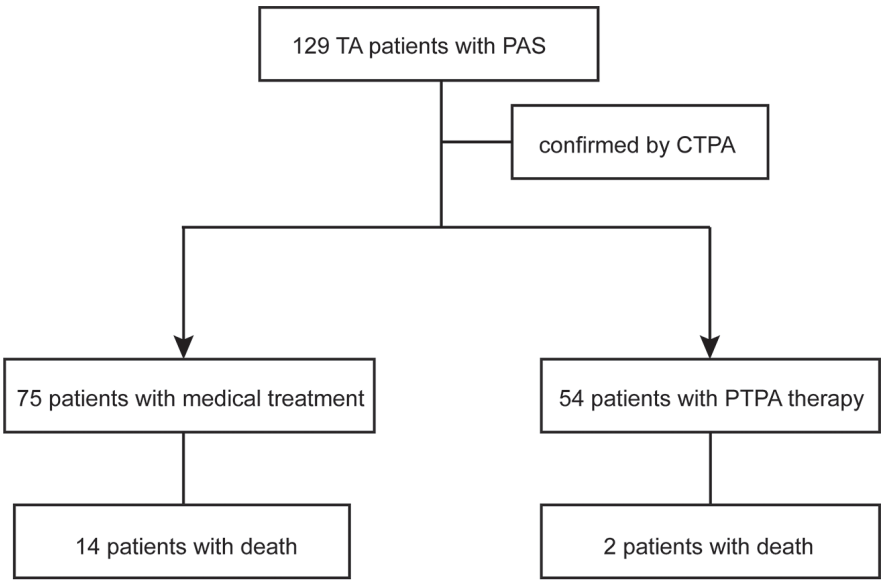

Fig. 1. Flow diagram of the study.
TA: Takayasu's arteritis; CTPA: computed tomographic pulmonary angiography; PAS: pulmonary artery stenosis; PTPA: percutaneous transluminal pulmonary angioplasty.

Statistical analyses

Continuous variables with normal distribution are expressed as mean with standard deviation, and continuous variables without normal distribution are expressed as median (interquartile range). Categorical variables are reported as absolute numbers and percentages. Differences between groups were tested using the independent t-test or Mann-Whitney U-test. Comparisons between groups were performed using the chi-square test for categorical data. Fisher's exact test was used if the expected frequencies were less than 5. Mortality rates were estimated using the Kaplan-Meier method, and differences between groups were assessed using the log-rank test. Test efficiency was set as the bilateral $\alpha = 0.05$, power $(1-\beta) = 0.8$. Statistical significance was set at a two-sided $p<0.05$. Statistical analyses were performed using the SPSS software (v. 25; SPSS, Chicago, IL, USA).

Results

Demographic data and clinical features

A total of 129 TAK patients (women, 101; men, 28; mean age, 40.5 years)

with PAS confirmed by CTPA and PH assessed using TTE were enrolled in this study (Fig. 1).

In the enrolled patients, 93 patients (72.1%) underwent RHC at baseline. 54 patients in the revascularisation group, the remaining is conservative drug treatment. Among these patients, 75 received conservative drug treatment and 54 underwent revascularisation (52 with PTPA and 2 with stent implantation). The median follow-up time was 54 (40.5–58.5) months. There were no significant differences in sex, age, comorbidities, disease activity, WHO function classification, 6MWD, and medication (except for endothelin receptor antagonists) between the groups. The patients' baseline clinical characteristics are shown in Table I.

Changes between groups at follow up

As shown in Table II, compared with the conservative treatment group, the WHO functional class and 6MWD significantly improved in the PTPA group ($p<0.001$ and $p=0.004$, respectively). The 6MWD obviously increased by 52 m in the PTPA group, while that in the control group only gained 29 m. And the level of N-terminal pro–B-type natriuretic peptide (NT-proBNP) was significantly different between the two groups ($p=0.005$), there was a clear downward trend in the PTPA group. There was also an apparent improvement in haemodynamic parameters assessed by TTE, including the right ventricular diameter (RVD), left ventricular diameter (LVD), tricuspid annular plane systolic excursion (TAPSE), and PASP ($p=0.016$, $p<0.0001$, $p<0.0001$, and $p=0.005$, respectively). Fourteen patients died in the conservative treatment group, as opposed to two patients in the endovascular group. All patients died from right-sided heart failure, which was confirmed by a death certificate. In terms of hard endpoints, cardiac death caused by TAK associated with PH was significantly different between the two groups ($p=0.031$, Fig. 2).

Complications of intervention

In the PTPA group, there were five (9.3%) patients with hematoma in the puncture point of the femoral vein. In addition, four patients (7.4%) had reperfusion pulmonary oedema (RPE) and three patients (5.6%) with haemoptysis during or after the intervention.

Table I. Clinical characteristics in TAK patients with pulmonary artery stenosis at baseline.

Variables	Medical therapy (n=75)	Endovascular treatment (n=54)	*p*
Clinical characteristics			
Age, years	40.6 ± 13.0	40.3 ± 13.0	0.878
Follow up, months	55.0 (43.0, 59.0)	48.5 (39.5, 58.0)	0.131
Female, n(%)	56 (74.7%)	45 (83.3%)	0.239
WHO FC I-II	22 (29.3%)	19 (35.2%)	0.481
WHO FC III-IV	53 (70.7%)	35 (64.8%)	0.481
6MWD	409 ± 88	426 ± 47	0.201
Comorbidities, n(%)			
Secondary hypertension	5 (6.7%)	7 (13.0%)	0.225
Dyslipidaemia	4 (5.3%)	5 (9.3%)	0.301
Diabetes mellitus	0 (0.0%)	0 (0.0%)	1.000
Smoking	2 (2.7%)	1 (1.9%)	0.622
CAD	0 (0.0%)	0 (0.0%)	1.000
PAD	28 (37.3%)	16 (29.6%)	0.363
Blood test			
CRP, mg/l	4.97 (2.65, 9.16)	3.18 (1.93, 4.94)	0.002
ESR, mm/h	5.0 (3.0, 7.0)	7.0 (2.0, 17.0)	0.034
TBIL, mmol/L	22.5 ± 15.3	19.3 ± 11.8	0.190
NTpro-BNP, pg/mL	754.3 (297.0, 2116.0)	435.0 (180.5, 1853.0)	0.098
Medications			
PDE-5 inhibitors	48 (64.0%)	32 (59.3%)	0.584
ERA	23 (30.7%)	30 (55.6%)	0.005
Prostacyclin Analogue	8 (10.7%)	7 (13.0%)	0.688
Prednisone	56 (74.7%)	42 (77.8%)	0.683
Immunosuppressants	6 (8.0%)	4 (7.4%)	0.901

Data are presented as the means ± SD, median or as numbers and percentages.
TAK: Takayasu's arteritis; WHO FC: World Health Organisation function classification; 6MWD: 6-min walk distance; CAD: coronary artery disease; PAD: peripheral arterial disease; CRP: C-reactive protein; ESR: erythrocyte sedimentation rate; TBIL: total bilirubin; NT-proBNP: N-terminal pro-brain natriuretic peptide; PDE-5: phosphodiesterase (PDE) 5.

Table II. The changes between medical therapy group and endovascular treatment group at follow-up.

Variables	Medical therapy (n=75)	Endovascular treatment (n=54)	*p*
Clinical changes			
WHO functional class, I/II/III/IV	25/26/3/0	14/39/16/6	<0.001
6MWD, m	438 ± 90	478 ± 48	0.004
NTpro-BNP, pg/mL	650.0 (146.4, 2063.0)	235.0 (124.5, 488.6)	0.005
ECHO			
RVD, mm	29.5 ± 7.2	27.0 ± 6.7	0.016
LVD, mm	39.0 ± 5.8	42.4 ± 4.5	<0.001
TAPSE, mm	17.2 ± 2.9	19.4± 2.6	<0.001
PASP, mm Hg	79.2 ± 26.3	62.0 ± 21.7	0.005
Cardiac death, n(%)	14 (18.7%)	2 (3.7%)	0.023

Data are presented as the means ± SD, median or as numbers and percentages.
TAK: Takayasu's arteritis; PH: pulmonary hypertension; 6MWD: 6-minute walk distance; RVD: right ventricular diameter; LVD: left ventricular diameter; TAPSE: tricuspid annular plane systolic excursion; PASP: pulmonary artery systolic pressure.

Discussion

The present study investigated the long-term outcomes of TAK patients with PAS and PH who received medical treatment or endovascular therapy. Under basic medicine, our outcome analysis indicates that PTPA therapy improves survival outcomes compared to medical treatment alone.

Previous reports have shown that pulmonary arterial involvement is frequent, occurring in 13.3%-61.7% of patients with TAK (5-8). However, pulmonary arteritis in the context of TAK is often overlooked in the clinical practice. The

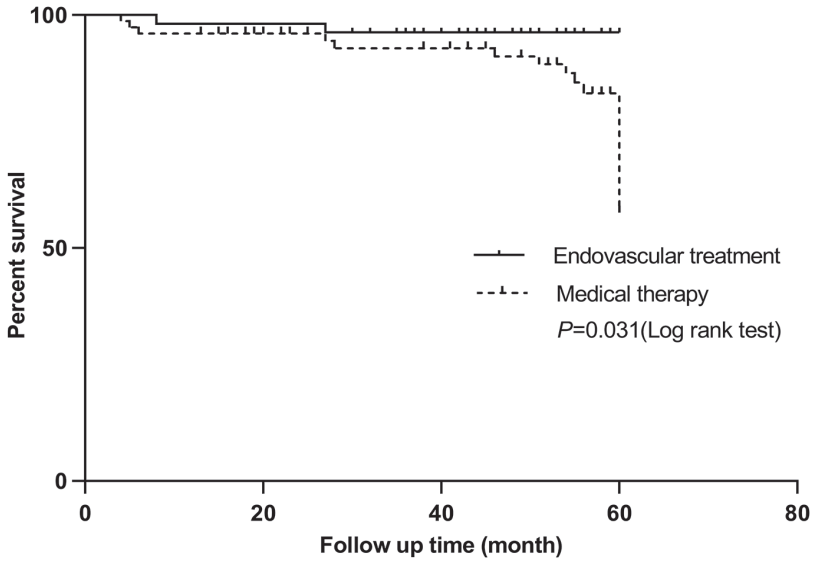

Fig. 2. Comparison of survival rate between patients with Takayasu's arteritis and pulmonary artery stenosis by medical treatment and percutaneous transluminal pulmonary angioplasty.

mechanism of TAK-related pulmonary arteritis remains unclear. Narrowing of the pulmonary arteries is mainly due to the extension of the inflammatory processes of intimal proliferation. Patients with PH due to TAK-associated PAS have a poor prognosis. Toledano *et al*. found that the mortality rate was 20.5% in patients with PAS and 33.3% in patients with PH (21).

However, there is currently no unified recommendation for the treatment of pulmonary artery lesions associated with TAK. Moreover, reports on long-term outcomes of TAK with pulmonary artery involvement are scarce. The treatment of TAK involving pulmonary vasculitis relies mainly on long-term drug therapy. Although a few case reports or small sample studies have found that PTPA treatment for TAK may be beneficial (13, 22), the relatively high intervention-related complications and mortality limit its widespread use. Feinstein *et al*. found that RPE was up to 61% after PTPA in 11 of 18 enrolled patients with RPE (23). Similarly, Kataoka *et al*. reported that 27 of 51 patients (53%) developed RPE (24). Postoperative RPE can be life-threatening, which is an important concern for interventional doctors.

Surprisingly, there was a significant reduction in the incidence of RPE with the introduction of the pressure wire approach during PTPA. Takumi *et al*. reported that the incidence of RPE was 6.9% after applying the pressure-wire technique (15). This study found that balloon dilation should be stopped to avoid RPE when the distal mean PAP, indicated by the pressure wire after each dilation, reached 35 mm Hg. Successful dilation was defined as a pressure ratio of distal to proximal pressures across the target lesion of ≥0.8, as detected by the pressure wire. Benefiting from the unique properties of the pressure wire, some interventional doctors began to use this technique with caution in patients with TAK and PAS. Zhou and colleague evaluated the results of 50 patients with TAK-PH who completed the PTPA procedure (the PTPA group) and 21 patients who refused the PTPA procedure (the non-PTPA group). The results indicated that there were 3 patients (6.0%) deaths occurred in the PTPA group and 6 patients (28.6%) in the non-PTPA group (P<0.05). PTPA tended to be associated with a reduced risk of all-cause mortality with acceptable safety and efficacy (17). After several trials, we believe that this technique makes PTPA safer for the treatment of TAK-associated PAS. Four patients (7.4%) developed RPE during or after the intervention, and all four recovered after treatment with diuretics, hormones, and non-invasive ventilators. The proportion of RPE in our study was similar to that reported in a previous study (15). Furthermore, we observed significant improvement in symptoms and exercise tolerance in patients treated with PTPA. Also, compared to medical treatment alone, the PTPA procedure improved the survival rate (P=0.031). It is widely accepted that PTPA is the preferred balloon dilation angioplasty for TAK-associated PAS, and our observations support this dogma. But two patients that received stent implantation for lesions located in a relatively big pulmonary artery and the results of balloon dilation were not fully satisfactory.

Conclusions

The present study has several limitations that need to be acknowledged. First, this was a retrospective study and the participating patients were enrolled from only two centres. Second, as this was a non-randomised controlled study, selection bias in the process of patient enrolment may exist. Also, the contribution of PH-targeted drugs to the improved survival cannot be ruled out. However, conducting prospective studies on this rare disease is very difficult. The advantage of this study is that it included a relatively large patient sample, and that these patients were followed up for a long period of time.

In conclusion, under basic medicine, PTPA therapy improves the long-term survival outcomes of TAK patients with PAS and PH, compared to medical treatment alone. PTPA may be a promising modality for the TAK patients with PAS and PH. Prospective, multicentre studies are needed to confirm the findings of our study in the future.

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