

# New endovascular techniques for the treatment of life-threatening Takayasu arteritis.

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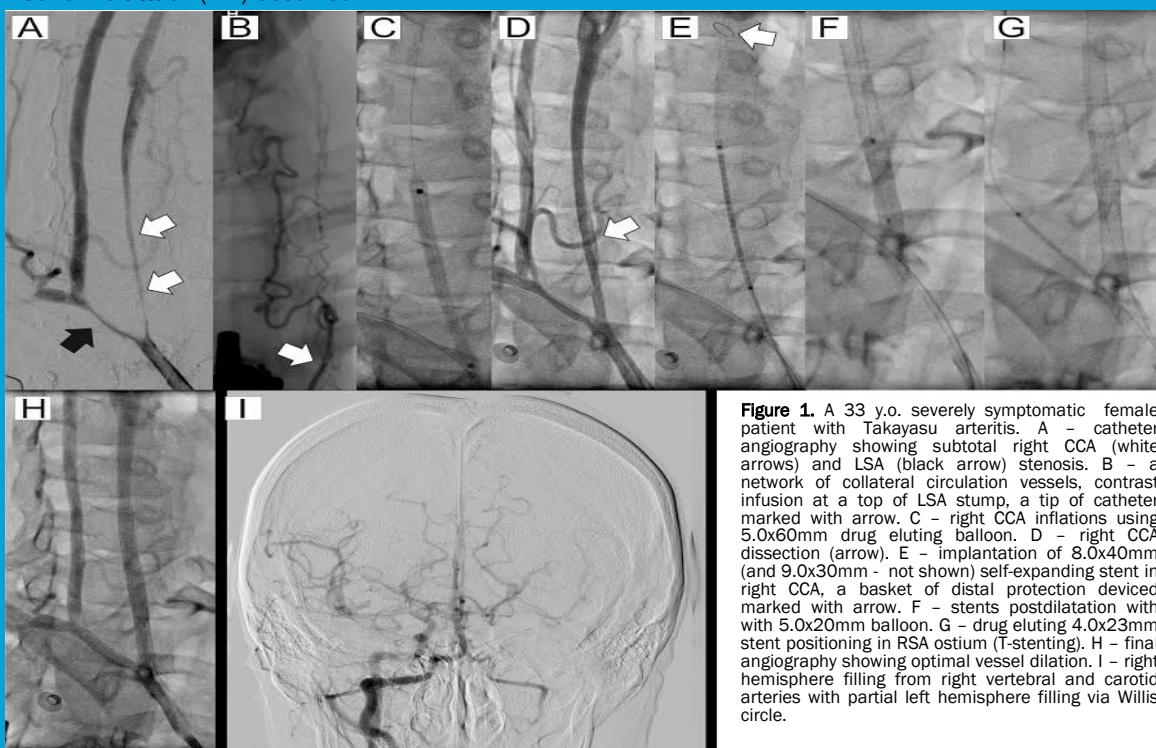
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## Introduction

Takayasu arthritis is a medium-size and large artery vasculitis of unknown aetiology affecting mainly aorta and its major branches, usually in young female patients. Histopathology reveals adventitial thickening, focal lymphocytic infiltration of tunica media and intimal hyperplasia leading to artery stenosis/occlusion. Less commonly media degeneration presents as aneurysmal dilatation [1]. Clinical symptoms arise from systemic inflammation and local vascular complications. Neurological manifestations include headache, dizziness, visual disturbance, TIA and stroke [2]. Takayasu arthritis may be associated with premature mortality among young patients. Mortality is significant (3-11%) and vary on geographical location and management strategy. Most cause of death includes stroke, myocardial infarction, congestive cardiac failure, peri- and postoperative complications. Majority of patients (23%) are unable to work, and about 60% are limited in everyday activities [3,4].

## Case report

We report a case of 33 y.o. lady being diagnosed on rheumatology ward in Jan 2014 due to unexplained weight loss, anemia and highly elevated systemic inflammation markers (C-reactive protein of 129 ng/ml, erythrocyte sedimentation rate of 107mm/h). Those findings were accompanied by severe dizziness and extremely low value of systemic blood pressure measured on arms. Doppler ultrasound (DUS) examination followed by computed tomography angiography (angio-CT; Mar 2014) revealed thickening of descending aorta wall, near-to-occlusion stenosis of both common carotid arteries (CCA), severe stenosis of right subclavian artery (RSA) and occlusion of left subclavian artery (LSA). According to the criteria of the large vessel vasculitis a diagnosis of Takayasu arthritis was made [2] and the patient was initiated on a combination therapy of cyclophosphamide, prednisolone and hydrocortisone. Within next 6 months, she was hospitalized several times due to persistent neurological symptoms including dizziness and drop attacks. In Dec 2014 clinical deterioration with increased neurological symptoms appeared. The patient was unable to adopt an upright position because of severe dizziness, moreover, right-hemisphere transient ischemic attack (TIA) occurred



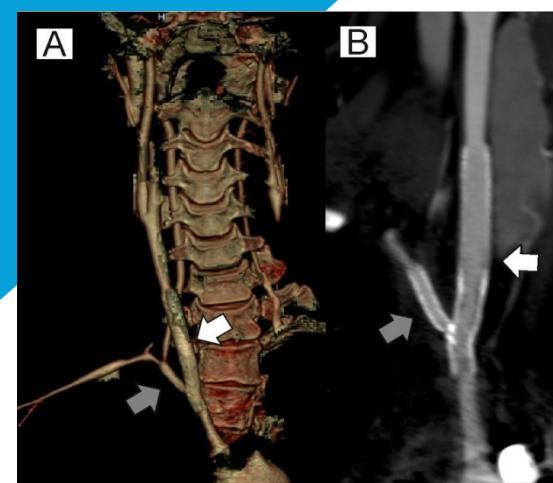
She was admitted to our Vascular Surgery Department in Jan 2015. After complex non-invasive diagnostic evaluation and multidisciplinary team (neurologist, vascular surgeon, cardiologist) consultation she was scheduled for invasive aortic arch angiography in terms of endovascular treatment. Via right femoral access, 6F diagnostics pig tail catheter was introduced. Angiography revealed: occlusion of LSA, occlusion of left CCA, significant narrowing of innominate artery (IA), long, 95% stenosis of right CCA, 80% stenosis of RSA. [Figure 1A]. Cerebral filling was extremely poor, mainly from collateral circulation [Figure 1B]. Right vertebral artery was the only patent vessel supplying brain. In accordance with ischemic symptoms and predicted procedure feasibility it was decided to perform an angioplasty of vessels supplying right hemisphere. On diagnostics Imager II Bern 5F catheter, V-18 Control Wire 0.0018in was navigated through IA to RSA. Diagnostic catheter was then changed to 40° angled 8F Mach1 Peripheral Guide Catheter (all above from Boston Scientific, Natick, MA, USA). Right CCA/ICA were wired with coronary BMW 0.014in wire (Abbott Vascular, Santa Clara, CA, USA) and predilatation of right CCA with 2.5x20mm coronary balloon catheter was performed (two inflation at 12atm and 14atm for 20sec each).

## References

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FilterWire EZ (Boston Scientific, Natick, MA, USA), a distal neuroprotection device was introduced and opened in right ICA.

Two inflations at 6atm/60sec using 5.0x60mm IN PACT Admiral Drug Eluting Balloon (DEB; Medtronic, Minneapolis, MN, USA) were performed in right CCA covering the whole stenotic segment [Figure 1C]. In spite of long-time inflations the patient complained only for temporary inflation-related right-side neck pain. As significant dissection of proximal and middle part of RCCA appeared [Figure 1D], two self-expandable stents (Precise PRO 8.0x40mm distally, 9.0x30mm proximally; Cordis Fremont, CA, USA) were implanted covering dissected segment as well as artery ostium and bifurcation with RSA. Postdilatation with 5.0x20mm balloon catheter (2x12atm/20sec) was done [Figure 1E,F]. The next target was RSA. Stent struts covering RSA ostium were crossed with 0.014in HT BMW 0.014in wire and then predilated with 2.0x20mm balloon catheter. Coronary, drug eluting stent (DES, Xience Pro 4.0x23mm; Abbott Vascular, Santa Clara, CA, USA) was implanted at 18atm/30sec and postdilated with 5.0 x20mm balloon catheter at 14atm/40sec. [Figure 1G]. A filter from RICA has been retrieved very easily. In control angiography correct location of stents with no residual stenosis and significant blood flow improvement to both hemispheres were recorded [Figure 1H,I]. Within next 12 hours significant improvement of general status and reduction of neurological symptoms were observed. The patient was discharged after 4 days of hospitalization on dual antiplatelet therapy (aspirin 75 mg/daily permanently and clopidogrel 75 mg/daily for six months). Control DUS performed 4 weeks later demonstrated good flow through implanted stents with no signs of restenosis. Three and eight months after the procedure, control angio-CT revealed all stents patency and no new stenotic lesions of treated vessels [Figure 2]. Up to now, patient has remained in good clinical status without any neurological disorders.



**Figure 2.** Control angio-CT of the patient performed 7 months after procedure. A - 3 D reconstruction showing all stents patency and no new stenotic lesions of treated vessels. B - 2D cross-section demonstrating undamaged right CCA (white arrow) and RSA (grey arrow) stents structure.

## Discussion

Surgical bypass arteries reconstruction has been shown to be superior to endovascular treatment [5]. However, this relates usually to lower limb artery atherosclerotic lesions. The higher risk of serious early and postprocedural surgical complications should be also considered [3,6,7]. With intensive technological development, endovascular treatment has been introduced as a alternative to surgery for arterial stenotic lesions in most of vascular territories. In TA, endovascular treatment has been found to be safe and very effective with acceptable 30-day complications rate of 7.1% in our cohort [8]. However, it has been shown that in-stent stenosis remains the main issue in both surgical and endovascular approach. [7,9]. Different mechanisms of artery stenosis development in non-atherosclerotic vasculitis, such as TA, including chronic intramural inflammation suggest that the use of drug-eluting systems might be an optimal way to deal with severe intimal hyperplasia leading to early and late restenosis affecting >30% of treated vessels [7,9,10].

Besides typical in-stent stenosis, external stent compression by progressive vessel wall fibrosis and calcification has been described [10]. However, it has been shown that in-stent stenosis remains the main issue in both surgical and endovascular approach. [7,9] Different mechanisms of artery stenosis development in non-atherosclerotic vasculitis, such as TA, including chronic intramural inflammation suggest that the use of drug-eluting systems might be an optimal way to deal with severe intimal hyperplasia leading to early and late restenosis affecting >30% of treated vessels [7,9,10]. Besides typical in-stent stenosis, external stent compression by progressive vessel wall fibrosis and calcification has been described [10]. In fact, we still miss the date of optimal endovascular treatment of TA, especially concerning drug-eluting devices use in supraaortic territory. Our case shows that such therapy may be safe and effective, also in middle-term observation.

We used DEB before stent implantation in RCCA because there has been no drug-eluting self-expanding stents available on market so far. This strategy gives a higher probability of restenosis avoidance. On the other hand, the use of drug-eluting stents has been shown to increase risk of early and late in-stent thrombosis, which in carotid territory might have devastating sequelae. The experience acquired in coronary interventions supports the strategy of at least 6 months dual antiplatelet therapy, as it was implemented in described case. Coherent systemic treatment with steroids and antiproliferative drugs is crucial in term of disease inhibition as drugs withdrawal might be associated with in-stent stenosis [11,12]

A very close follow-up observation may not be mandatory after atherosclerosis-origin lesion angioplasty. For carotid territory, self-expanding stent implantation is associated with 5-7% risk of significant in-stent stenosis and it is observed mainly during first year of follow-up [13]. In TA the risk of restenosis is significantly higher reaching >50% at 5 years and probably it does not decrease with time [8]. The risk of symptomatic disease progression related to new stenotic lesion is also very high and exceed 70% at 5 years [8,14]. Those findings support watchfull and frequent DUS and angio-CT examination that have been performed in our patient. As DUS is not a perfect image modality for proximal segments of aortic arch arteries evaluation thus it was decided to carry out regular angio-CT scanning. This examination enables optimal visualisation of all supraortic arteries including not only vessel wall but also stent structure in terms of potential stent deformation/fracture.

Outcomes of vascular intervention in TA may be improved by detailed preoperative assessment including measurement of disease activity, and by ensuring optimal immunomodulatory therapy before and after the procedure [12].