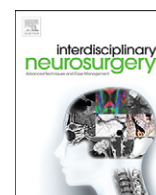


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Case Reports &amp; Case Series (CRP)

## Extracranial–intracranial bypass for Takayasu's arteritis: A case report

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### ABSTRACT

**Objective:** Takayasu's arteritis is a rare disease that often affects the great vessels at the origin as they branch from the aortic arch. There are numerous case series and case reports in the cardiothoracic literature describing carotid bypass to reestablish blood flow past the areas of stenosis. We present a rare case of thrombus forming in the distal ICA and occluding intracranial vessels, thus necessitating neurosurgical evaluation and intervention.

**Methods:** This is a case report of a 21-year-old right-handed female who presented with right-sided hemiplegia, facial weakness, and language impairment. Radiological imaging showed occlusion of the left middle cerebral artery (MCA) and bilateral distal carotids. MR angiogram of the head and neck confirmed the above findings and also showed high-grade stenosis of the right vertebral artery at its origin. An EC–IC bypass was necessary to maintain perfusion to the left side of her cortex and to prevent further neurological deficits.

**Results:** At the 9 month follow-up, her overall neurological exam has improved from complete paralysis on the right side to more than antigravity in her motor strength assessment. The follow-up imaging showed that the bypass remained patent while the ICA remained occluded.

**Conclusion:** STA–MCA bypass to enhance cerebral blood flow in the setting of stroke symptoms due to Takayasu's arteritis can serve as a bridge before definitive cardiothoracic treatment of the great vessels.

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### Introduction

Takayasu's arteritis is a rare vasculitis that affects the great vessels branching off the aortic arch. It is commonly seen in the Asian descent population with a female predominance ranging from 3:1 to 8:1 [1]. The initial presenting symptom is often pulselessness in the upper extremities and in severe cases, stroke-like symptoms [2]. Current medical treatment options include corticosteroid, immunosuppressive agents, and anti-TNF therapy. More invasive/surgical options include percutaneous transluminal angioplasty (PTA) or surgical vascular reconstruction.

It is rare for Takayasu's arteritis to affect the intracranial vessels, however there have been reports of the inflammatory arteritis causing narrowing of the intracranial vessels resulting in strokes [2–4]. With the lumen of the carotid and/or vertebral arteries narrowed by the disease process neurological deterioration ensues due to inadequate intracranial blood supply.

In the event of inadequate cerebral blood flow, an extracranial–intracranial (EC–IC) bypass should be considered in appropriate cases to augment the cerebral metabolic demand. This report demonstrates the

safe and successful use of the superficial temporal artery-to-middle cerebral artery (STA–MCA) bypass in the setting of Takayasu's arteritis.

### Case report

A 21-year-old right-handed, previously healthy, female initially presented with a right-sided hemiplegia, dysarthria, and facial weakness. Upon taking her vitals, it was noted that a blood pressure was unattainable from her right arm along with an absence of brachial pulse to palpation. The patient stated that she has been told by other medical practitioners that they were unable to get a blood pressure from her right arm. With further questioning, she stated that over the past 2 months her right arm had become clumsier and she had also been dropping things with extended use of her arm. Furthermore, at the same time she complained of right sided neck and jaw pain that was diagnosed as temporomandibular joint disorder by a dentist.

A non-contrast CT of the head performed at an outside hospital, prior to transfer, showed a hyperdense MCA sign that prompted immediate transfer to our hospital. A CT angiogram revealed a left middle cerebral artery (MCA) and bilateral distal internal carotid occlusion. She underwent tPA administration for a thrombus at the left ICA terminus with improvement in her right-sided strength, but only transiently. She developed a dense right hemiparesis and fluctuating dysphasia. Vasopressor treatment initiated by the critical care team improved her speech and stabilized her exam. Over the subsequent days attempts to wean her from her vasopressor

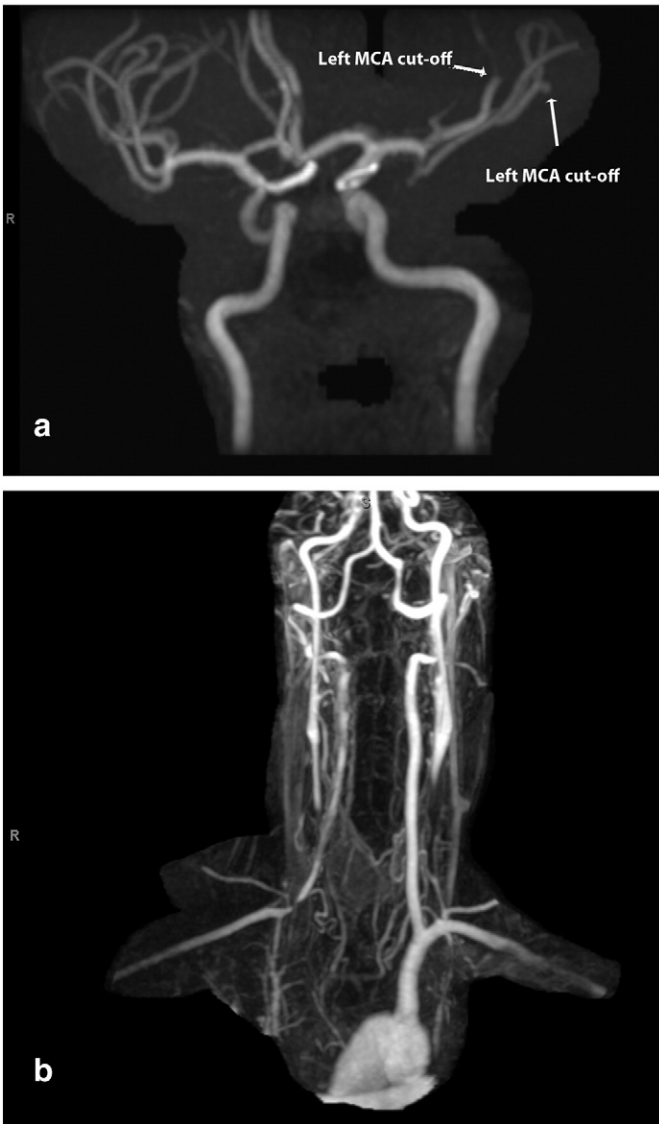
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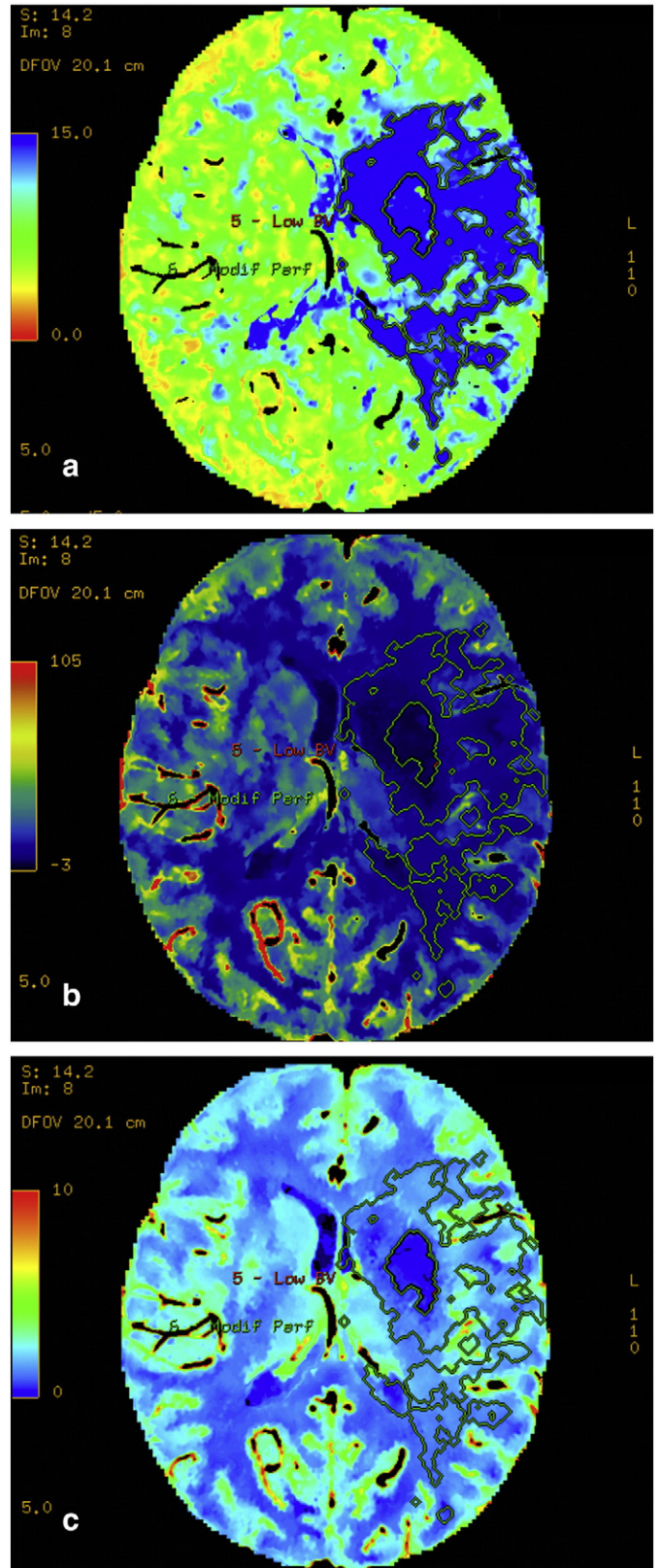
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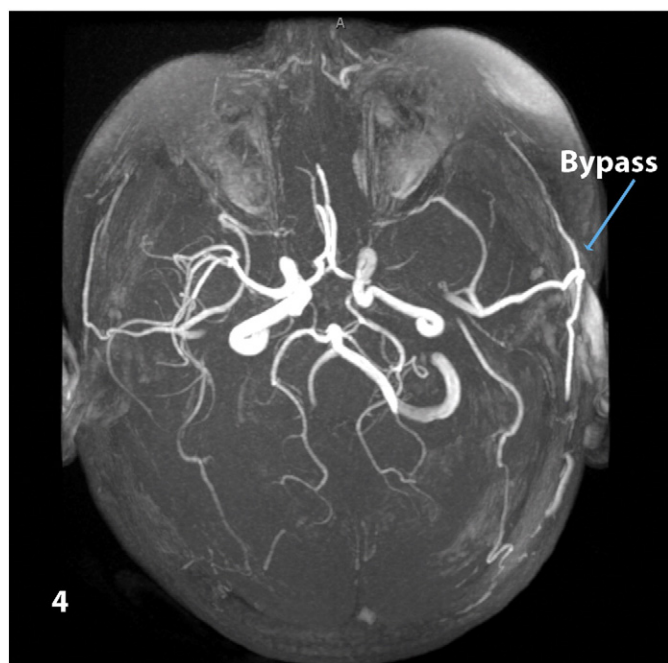


**Fig. 1.** Initial MR study after tPA administration, a) MRA of the head showing narrowing and peripheral cut-off of the proximal left MCA branches with b) MRA reconstruction of the neck demonstrating dissection of both common carotid arteries, innominate artery and right subclavian artery.

requirement were met with recurrent speech deficits. Neurosurgery was consulted at that point. MR angiogram of the head and neck confirmed the findings above and also showed high-grade stenosis of the right vertebral artery at its origin (Fig. 1). Cerebral blood flow studies (Fig. 2) were used and it was ultimately determined that the patient required an EC-IC bypass in order to preserve her language and speech. The imaging also demonstrated reconstitution of the left external carotid artery via small superficial collaterals with an adequate caliber STA to consider bypass. She underwent left STA-to-M4 branch of the left MCA end-to-side anastomosis with careful attention to maintenance of her blood pressure during the procedure by the anesthesia providers. The STA vessel was noted to be quite fragile compared to a more normal STA, but this did not preclude safe handling of the vessel or performance of the anastomosis. Post-operatively her vasopressors were weaned and discontinued without speech disturbance or deterioration in her hemiparesis.



**Fig. 2.** CT perfusion study showing a) Mean transit time, b) blood flow, and c) blood volume showing ischemic penumbra in the MCA territory with an infarcted core in the basal ganglia.



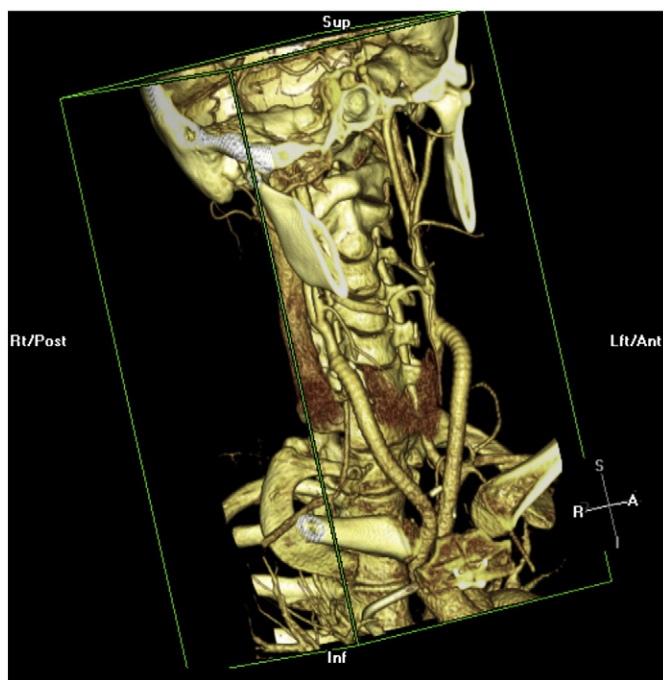
**Fig. 3.** MRA head reconstruction showing patency of left STA–MCA bypass at 9 month follow-up.

We obtained a follow-up MR angiogram at her 9 month clinic visit which continued to show the occlusion in the proximal MCA segment with filling of the M2 and distal vessels supplemented by the STA–MCA bypass (Fig. 3). Before this visit, she had followed-up with the cardiothoracic surgeons and had a bilateral carotid bypass performed without complication (Fig. 4).

Her neurological exam improved from a complete right-sided hemiplegia at initial consultation to a motor strength of 3/5 in her right upper extremity and 4/5 strength in her right lower extremity at 9 month follow-up. She has remained on an aspirin 325 mg regimen since her STA–MCA bypass and will continue it indefinitely.

## Discussion

Takayasu's arteritis is a rare disease process that is often encountered by rheumatologist and in surgical cases by the thoracic/vascular surgeons. Many times surgical intervention is required if the disease is severe enough or if treatment is refractory to best medical management. In this case, our patient demonstrated 5 of 6 criteria for diagnosing Takayasu's arteritis with a sensitivity of 92.1% and specificity of 97% [5]. The 1990 criteria for classification include: 1) Age onset <40 years 2) Claudication of extremities (including worsening fatigue, discomfort in muscles while in use in the upper extremities) 3) Decrease brachial artery pulse 4) Blood pressure difference > 10 mmHg 5) Bruit over subclavian arteries or aorta 6) Arteriogram abnormality. Our patient exhibited all of these criteria except the bruit. A more invasive cerebral angiography was considered, however it was not performed in order to avoid the risk of vessel lumen intimal damage and we felt that it would not have changed the patient's diagnosis and/or management. Other diagnostic procedures, such as a vessel biopsy would have been low yield since it was obvious that our patient had demonstrated that she required a surgical intervention of her *great vessels*. This situation, however, was complicated by the fact that she was having strokes due to an occlusion of her left MCA. The decision was made to perform the STA–MCA bypass first in order to stabilize her neurological condition.



**Fig. 4.** CT neck 3-D reconstruction after bilateral carotid artery bypass.

She was unable to tolerate a decrease in her systolic blood pressure without developing aphasia. Her hemiplegia persisted despite alterations in increasing her cerebral perfusion medically. The CT perfusion scan did confirm inadequate cerebral blood flow to the left MCA distribution (Fig. 2). After the STA–MCA bypass her neurological symptoms stabilized without the use of vasopressor medications.

STA–MCA bypass is routinely used in cases of moyamoya disease [6] or as a bypass for complex aneurysms that would otherwise cause a complete occlusion of the distal MCA distribution. It is our belief that in cases of intracranial vessel occlusion from Takayasu's arteritis an EC–IC bypass can be considered in appropriately evaluated patients.

## Conclusion

We have demonstrated that the STA–MCA bypass is a versatile procedure that is effective in directly augmenting cerebral blood flow to meet metabolic demand in the setting of Takayasu's arteritis. The bypass served as a safe and effective bridge to successfully wean the patient from vasopressors and until definitive reconstruction of the great vessels could be performed. Even after that was achieved the bypass remained patent and supplemented collateral flow to the occluded territory of the MCA.

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