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One sided bypass for bilateral Moyamoya disease, a case report and review of the literatures

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Keywords: cerebral angiography; intraventricular haemorrhage; Moyamoya disease; vascular anastomosis

Abbreviations: CBF, cerebral blood flow; CT, computed tomography; CTP, computed tomography perfusion; DSA, digital subtraction angiography; ICA, internal carotid artery; ECA, external carotid artery; MCA, middle cerebral artery; MMD, Moyamoya disease; MRI, Magnetic resonance imaging; STA, superficial temporal artery.

Abstract

Background: Moyamoya disease (MMD) is a rare condition, where the most appropriate treatment for it is yet to be determined. Surgery remains an important method of choice although it is considered a form of palliative care. The outcome following surgery is very difficult to judge, and there is no standardised measurement to assess it. It is therefore important to know which approach for such patient is adequate.

Clinical Presentation: A 21-year-old male patient presented with signs and symptoms of intracranial haemorrhage. Upon investigation, a diagnosis of bilateral MMD was made, and one-sided direct bypass surgery was subsequently performed. At 3-year follow-up, there is no evidence of recurrent cerebral vascular event.

Conclusion: This case provided further evidence that direct bypass surgery is beneficial for patient in terms of blood flow improvement and symptom relieve. Although there is no consensus on whether bilateral surgical intervention is mandatory for patient with bilateral MMD, unilateral bypass might be sufficient enough. Further study is required to evaluate the best approach for such group of patient.

Introduction

Moyamoya disease (MMD) was first reported in 1957 by Shimizu and colleagues as hypoplasia of the bilateral internal carotid arteries (ICAs). Moyamoya syndrome has been used to describe a similar condition when an underlying pathology, such as atherosclerosis, radiation therapy or sickle cell disease, can be identified. The syndrome consists of unilateral or bilateral steno-occlusive arterial changes and associated Moyamoya collaterals [1]. The incidence of MMD peaks in two age groups: children who are approximately 5 years of age and adults in their mid-40s [2]. Chances of having MMD are 2-folded higher in female than male [2]. Three types of research have been conducted to explain the pathogenesis of MMD: pathological analysis of affected tissue, genetic-linkage studies and studies of the role of angiogenesis and extracellular matrix–related peptides in disease development and progression. Pathological analysis has revealed that vessel occlusion is not due to arteriosclerotic or inflammatory, but is a result of smooth-muscle cell hyperplasia and luminal thrombosis [3]. Suzuki and Takaku described six separate angiographic stages of Moyamoya based on the pattern of steno-occlusion and collateral formation.[4] Grade I refers to the narrowing of the ICA apex without Moyamoya collaterals. Grade II refers to ICA stenosis along with initiation of Moyamoya collaterals. Grade III refers to progression of the ICA stenosis with intensification of Moyamoya collaterals. Grade IV refers to development of external carotid artery (ECA) collaterals. Grade V refers to intensification of ECA collaterals along with a reduction of Moyamoya collaterals. Grade VI represents the final stage of the disease process with total occlusion of the ICA and disappearance of Moyamoya collaterals [4]. The pathogenesis of MMD is irreversible despite treatment. Current available treatments are aiming to prevent

stroke by improving cerebral blood flow (CBF) to the affected cerebral hemisphere. Medical therapy has been used in patients with MMD, particularly in mild cases or where surgical intervention is contraindicated. But there is few data showing either its short-term or long-term efficacy. Surgical treatment for patients with MMD typically uses the ECA as a source of new blood supply to the ischaemic hemisphere. There are two types of revascularisation: direct and indirect. In direct revascularisation, a branch of the ECA is directly anastomosed to a cortical artery. In indirect revascularisation, the ECA in direct contact with the brain, leading to an ingrowth of new blood vessels to the underlying cerebral cortex [3].

Case report

A 21-year-old male presented with a history of repeated headache accompanied by vomiting for 5 days. His memory and learning ability are worse than his peers. His past history and family history are unremarkable. On examination, he was afebrile and conscious There is no papilledema or other neurological deficit. Computed tomography (CT) of the head showed bilateral intraventricular haemorrhage. Diagnostic digital subtraction angiography (DSA) confirmed stenosis at the bifurcation of right ICA and anterior cerebral artery with collateral vessels (Figure 1) . The left ICA and the anterior cerebral artery are partially occluded with collateral vessel formation (Figure 1). CT angiography (CTA) of the head was also performed with similar findings as the DSA (Figure 2). CT perfusion (CTP) of the head revealed a hypovolemic left cerebral hemisphere comparing to the right (Figure 2).

The diagnosis of bilateral MMD was established based on the above evidence, with a more severe imagine findings on the left. Diameters of the following arteries are measured by vascular ultrasonography, anterior branch of left superficial temporal artery (STA) (1.4mm), posterior branch of STA (2.0mm). The anterior branch of the right STA is missing and posterior branch of STA (1.5mm). The reasonable vascular diameters improved the chances of success anastomosis. Since the lesion on the left side is more severe than on the right side (Figure 1 and 2). A decision to perform a unilateral bypass surgery on the left side was made. The procedure of STA to middle cerebral artery (MCA) bypass was carried out giving its clinical outcomes, particularly in diminishing the incidence of recurrent ischaemic events. The patient underwent an uneventfully operation with no postoperative complications. The patient was fit to be

discharged on the 14th post-operative day. CTA 2 weeks post-operatively showed patent vascular anastomosis with newborn compensatory proliferated vasculature leading to improved CBF as demonstrated by the CTP (Figure 2). One month post-operative DSA also demonstrated anastomosis patency (Figure 1). At 3-year follow-up, there is no evidence of intracranial haemorrhage or other complication. Unfortunately, no more image follow-up is feasible after one month since the patient is from a distant rural area.

Discussion

We presented a unilateral surgical intervention for a 21-year-old male with bilateral symptomatic MMD by STA-MCA bypass. The direct approach by STA-MCA bypass is a reliable treatment for MMD, which can immediately improve symptoms and reduce recurrent ischaemic events. Therefore it is the procedure of choice for this patient. Although a favourable clinical outcome is observed in this case, this may not necessarily match the improvement in the haemodynamic parameters. In addition, although no evidence of long-term complication, whether this is a result of the revascularisation is unknown. Especially when there is no long-term DSA data available in this case.

The STA-MCA procedure is technically demanding and can be associated with devastating haemorrhage. Usually, direct bypass is limited to adults or elder children due to the small caliber of the STA. Indirect bypass can also be considered in patients with serious medical comorbidities or in patients with inadequate recipient or donor artery grafts. Encephalomyosynangiosis, for example, is a safe and easy procedure, which can be offered in these groups of patients. However, this approach also has its disadvantages. Firstly, it requires larger craniotomy with dura opening. Secondly, it is associated with significant postoperative complications such as seizures and/or mass effect on the brain. Thirdly, it requires longer time to establish collateral vessel formation [5].

In recent years, surgical revascularisation is recommended in most symptomatic cases to reduce ischemic symptoms and to improve hemodynamic status [6]. Direct

revascularisation in particular, provides significant improvement in this scenario [7]. However, the effect on CBF after revascularisation remains controversial. In one study, short-term follow up after 3-6 months showed decreased CBF after direct revascularisation at the ipsilateral symptomatic hemisphere [7]. In other study, total CBF improvement can be achieved after STA–MCA bypass in symptomatic patients [8]. Evidence also showed that unilateral revascularisation could increase CBF in the contralateral non-intervened hemisphere [9]. In one study, Chen *et al* demonstrated significantly improved CBF in less than 2 weeks after operation [10]. CTP was used to in this study to provide a quantitative analysis of CBF before and after STA-MCA anastomosis [10].

The most appropriate revascularisation procedure for patients with MMD is not yet well defined. Nonetheless, ample evidence suggests that surgical intervention improves the outcome of patients with symptomatic MMD. Each procedure has its own advantages and disadvantages. Direct revascularisation may lead to immediate improvement of symptoms and could decrease recurrent ischaemic events. Although technically demanding and the possibility of life-threatening haemorrhage, direct bypass remains the preferred procedure for patients with symptomatic MMD. Previously, researchers have not used standardised outcome measures to assess patients on presentation and on long-term follow-up. It is suggested that neuropsychological testing after surgical intervention is critical to accurately assess outcomes [11]. Therefore, it should be integrated into future clinical trials. Direct and indirect revascularisation will continue to play a major role in the treatment of MMD. The choice of procedure appears to depend on the surgeon's experience and on the

nature of the patient's presentation. Randomised prospective clinical trials comparing the various procedures may be developed to define the most appropriate indications in different patient groups. Although beneficial presentation is evident in our patient after the unilateral STA-MCA bypass, no solid conclusion can be drawn from a single case. However, this paper opened an argument and also added one piece of evidence that unilateral direct bypass might be sufficient for patient with bilateral MMD.

Please state any conflicts of interest

All authors must disclose any financial and personal relationships with other people or organisations that could inappropriately influence (bias) their work. Examples of potential conflicts of interest include employment, consultancies, stock ownership, honoraria, paid expert testimony, patent applications/registrations, and grants or other funding.

Nothing to declare

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All sources of funding should be declared as an acknowledgement at the end of the text. Authors should declare the role of study sponsors, if any, in the collection, analysis and interpretation of data; in the writing of the manuscript; and in the decision to submit the manuscript for publication. If the study sponsors had no such involvement, the authors should

so state.

No

Ethical Approval

Research studies involving patients require ethical approval. Please state whether approval has been given, name the relevant ethics committee and the state the reference number for their judgement.

This is a retrospective case report. No ethical approval is required.

Consent

Studies on patients or volunteers require ethics committee approval and fully informed written consent which should be documented in the paper.

Authors must obtain written and signed consent to publish a case report from the patient (or, where applicable, the patient's guardian or next of kin) prior to submission. We ask Authors to confirm as part of the submission process that such consent has been obtained, and the manuscript must include a statement to this effect in a consent section at the end of the manuscript, as follows: "Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request".

Patients have a right to privacy. Patients' and volunteers' names, initials, or hospital numbers should not be used. Images of patients or volunteers should not be used unless the information is essential for scientific purposes and explicit permission has been given as part of the consent. If such consent is made subject to any conditions, **the Editor in Chief** must be made aware of all such conditions.

Even where consent has been given, identifying details should be omitted if they are not essential. If identifying characteristics are altered to protect anonymity, such as in genetic pedigrees, authors should provide assurance that alterations do not distort scientific meaning and editors should so note.

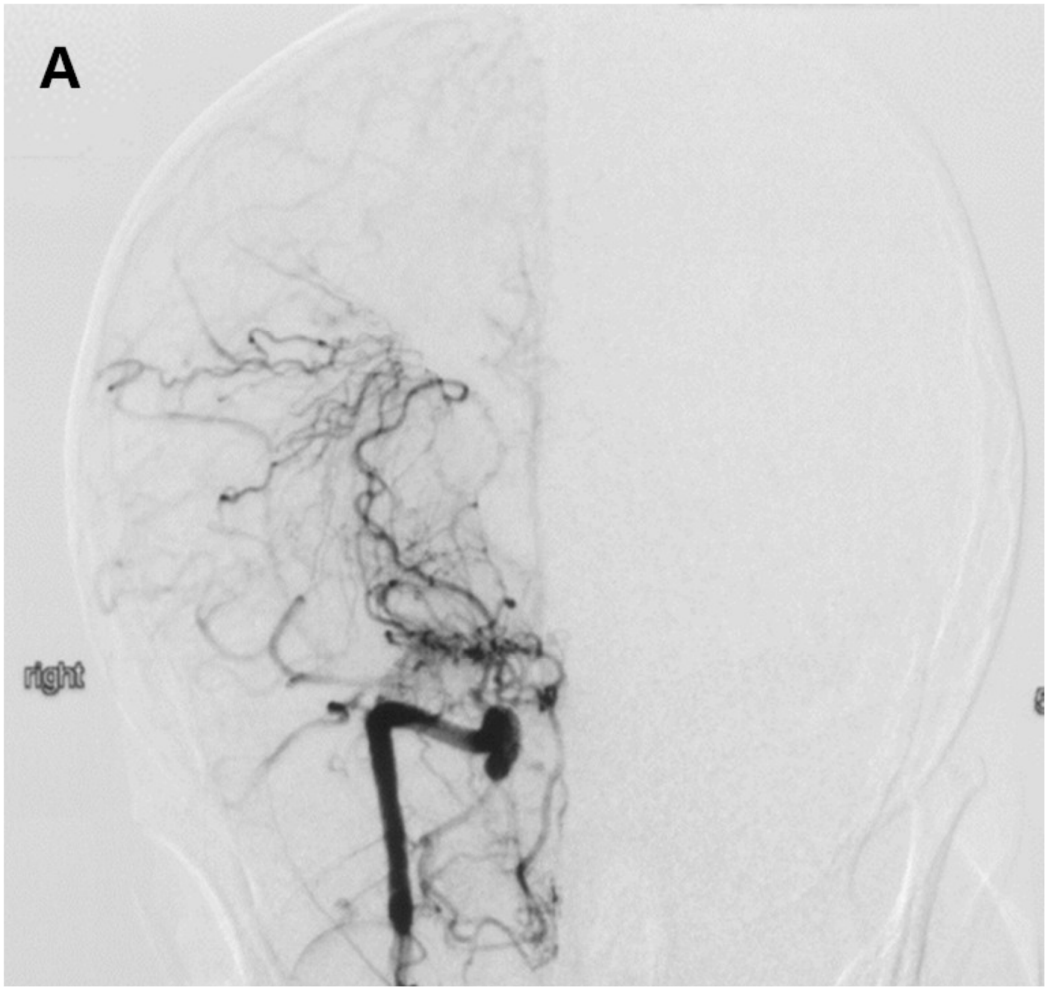
Written patient consent for publication has been obtained.

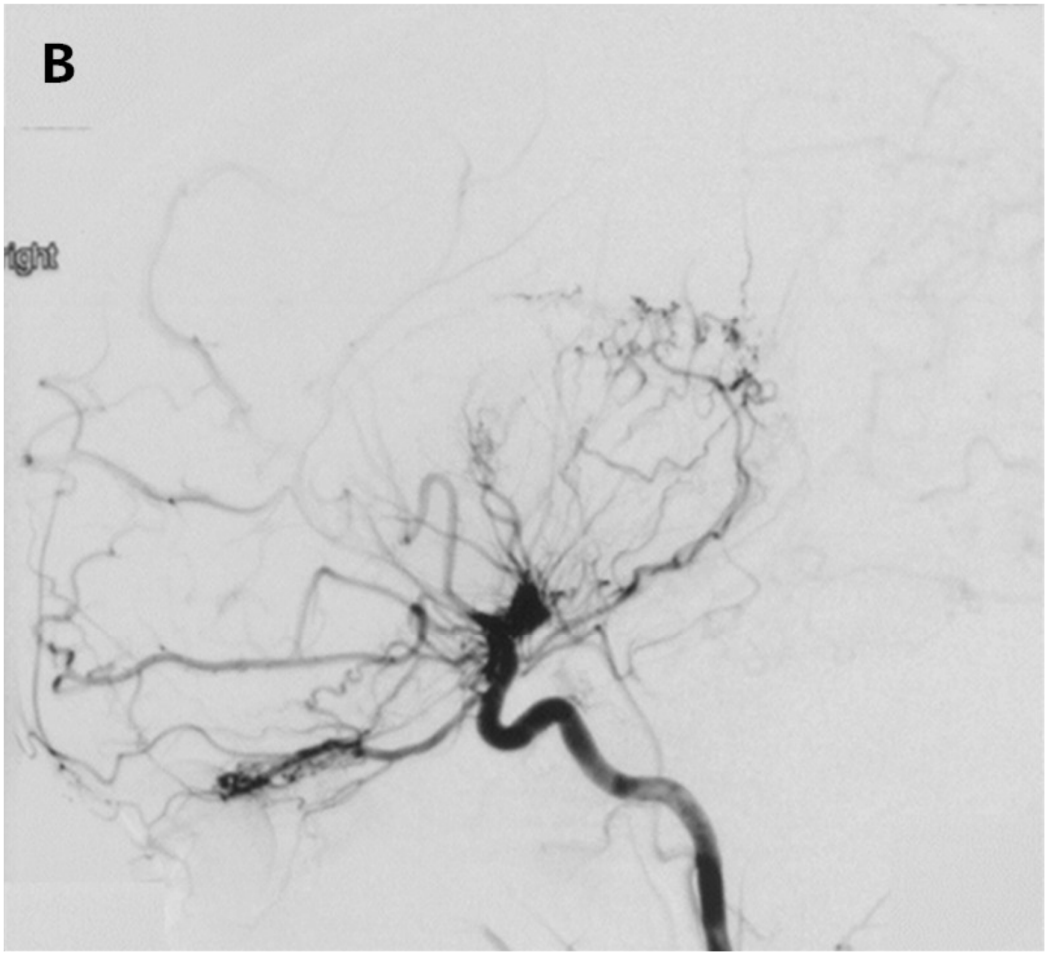
Author contribution

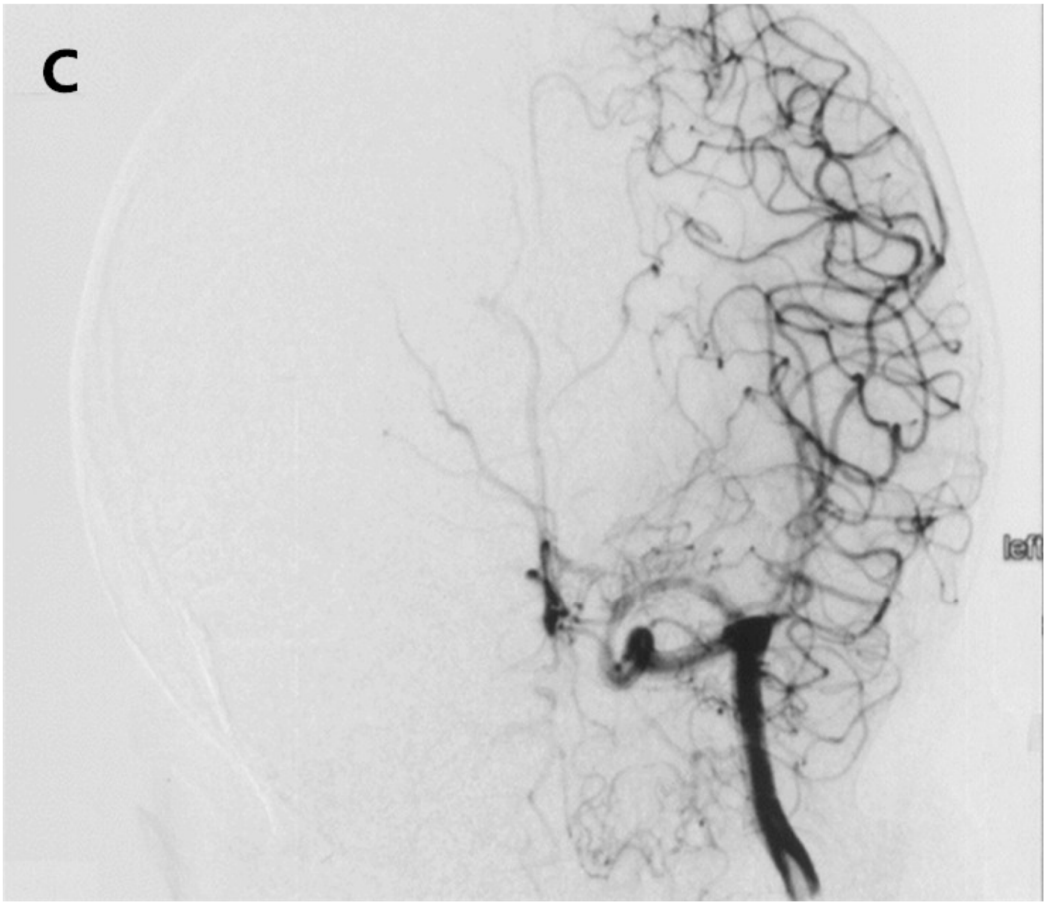
Please specify the contribution of each author to the paper, e.g. study concept or design, data collection, data analysis or interpretation, writing the paper, others, who have contributed in other ways, should be listed as contributors.

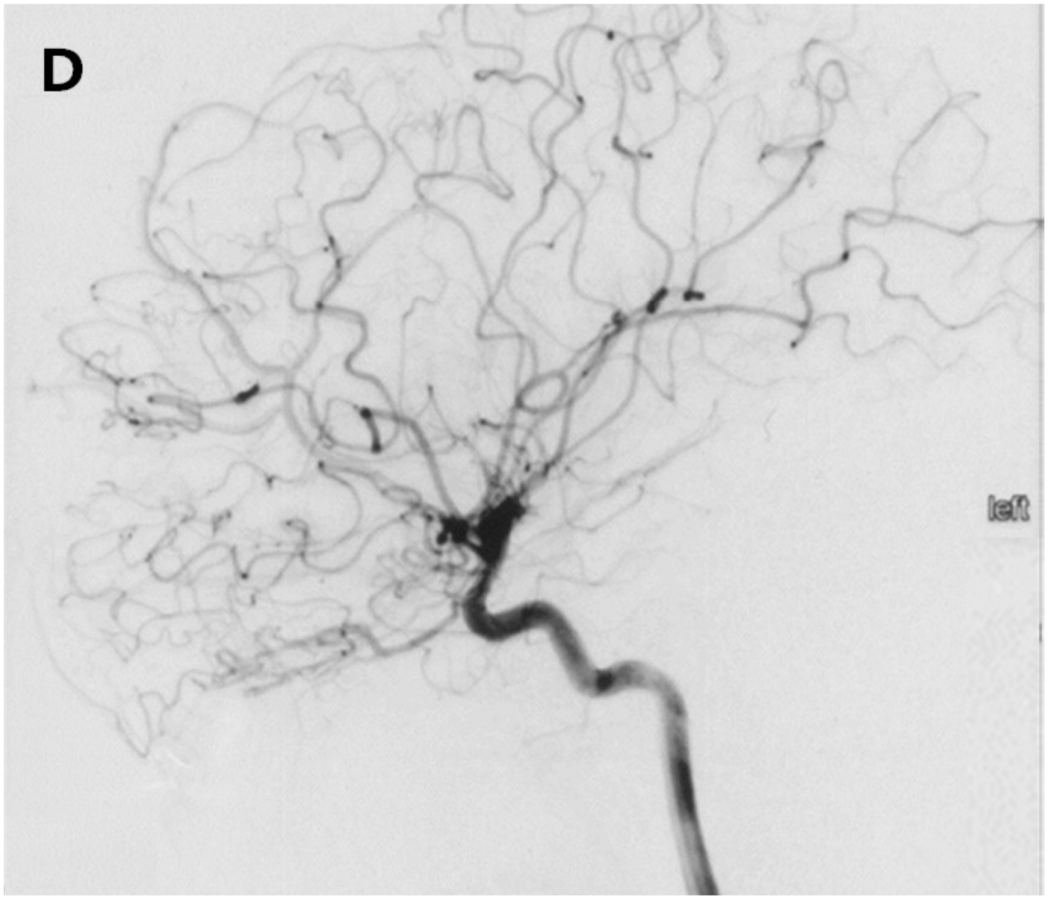
XL and NZ performed the surgery and participated in patient care; XL retrieved clinical data; XL and ZY reviewed the literature and wrote the paper.

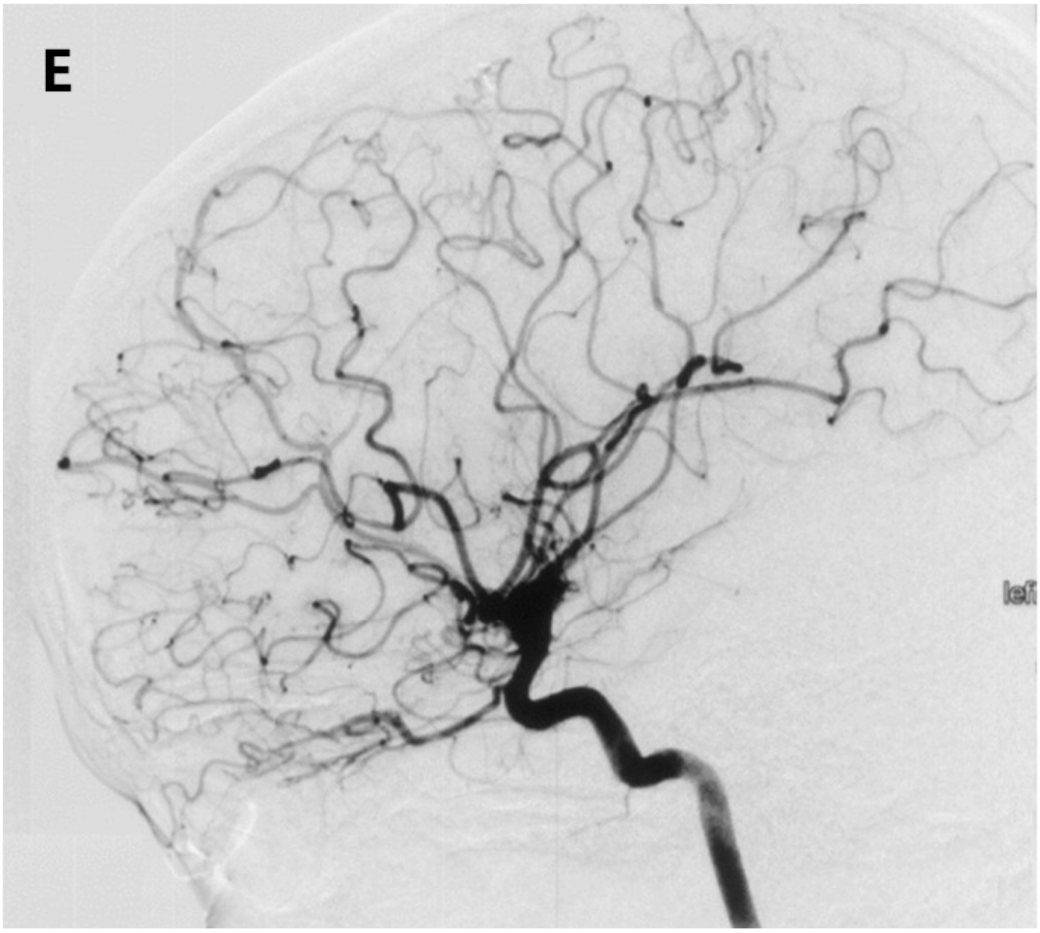
Figure 1. Digital subtraction angiography (DSA): pre-operative DSA showed (A-B) stenosis at the bifurcation of right ICA and anterior cerebral artery with collateral vessels, (A) Towne's view, (B) lateral view. Pre-operative DSA showered (C-D) partially occluded left ICA and anterior cerebral artery with collateral vessel formation, (C) Towne's view, (D) lateral view. One month post-operative DSA showed development of leptomeningeal anastomosis on the left side (E, lateral view) and right side (F, lateral view).











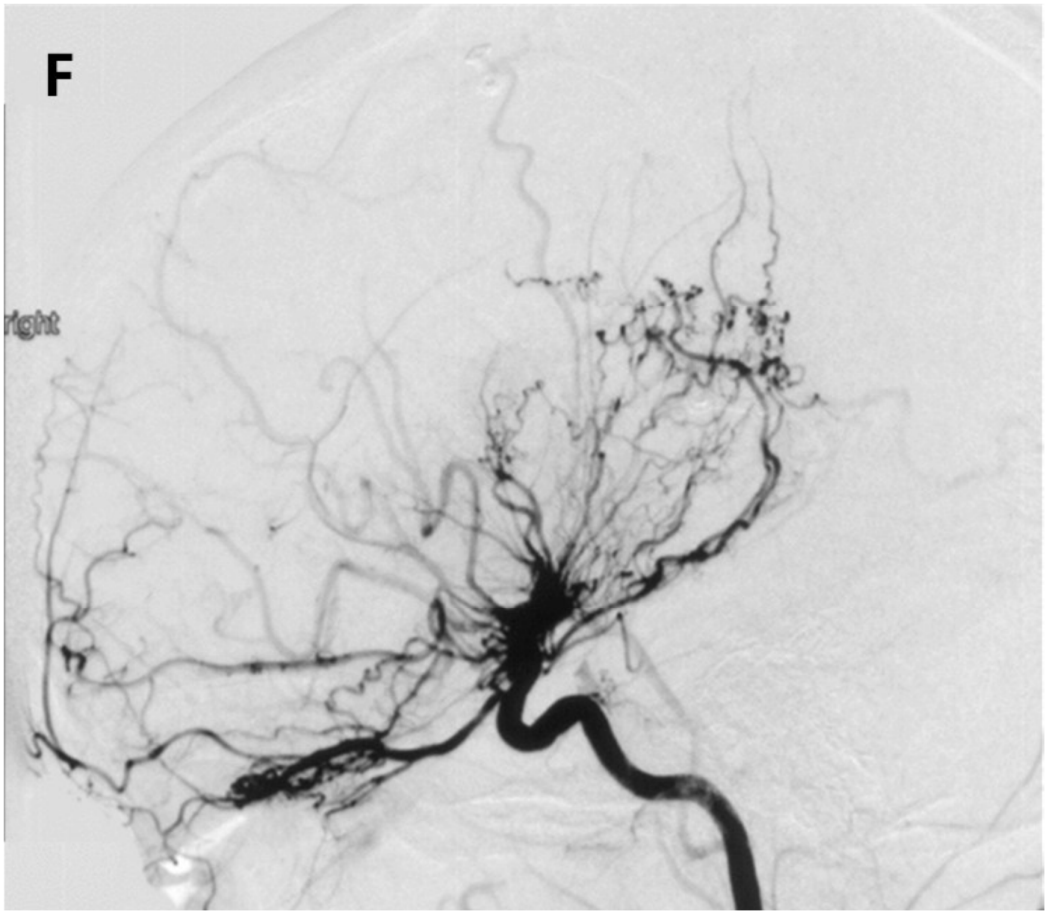
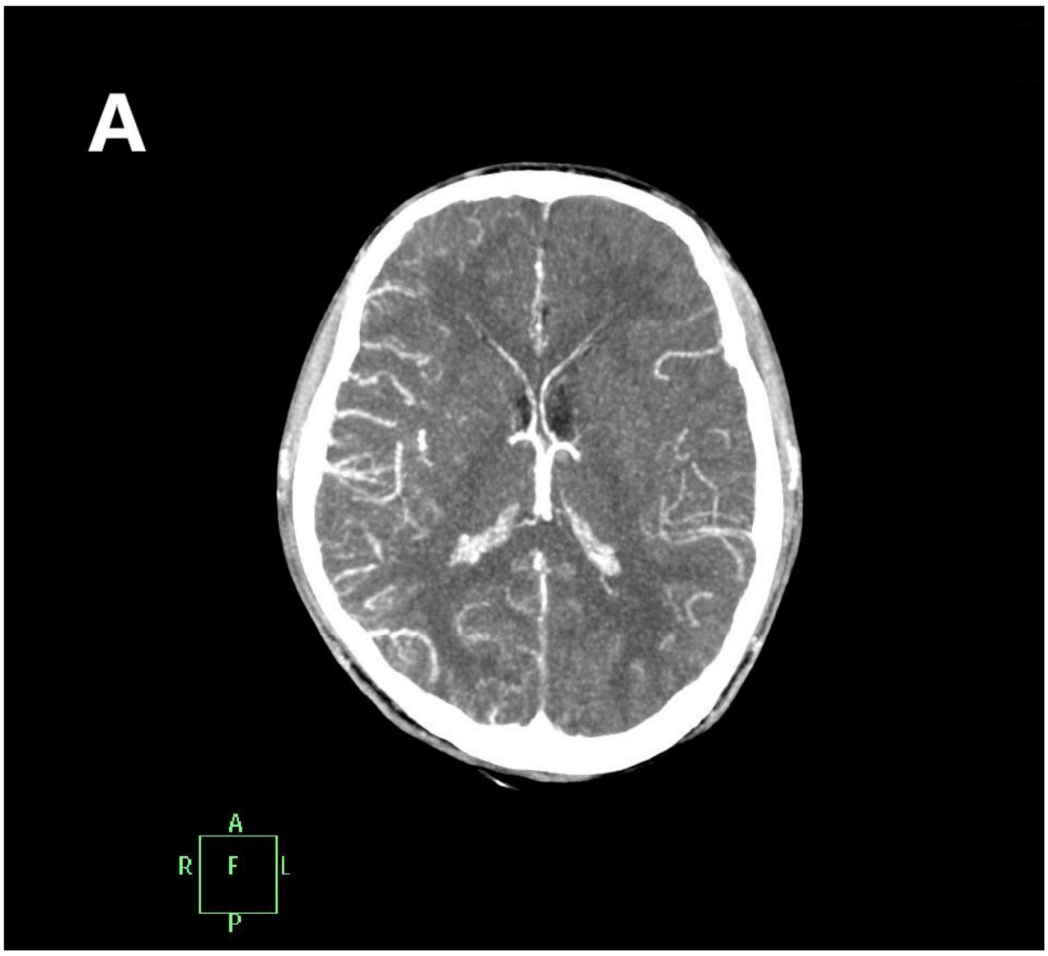
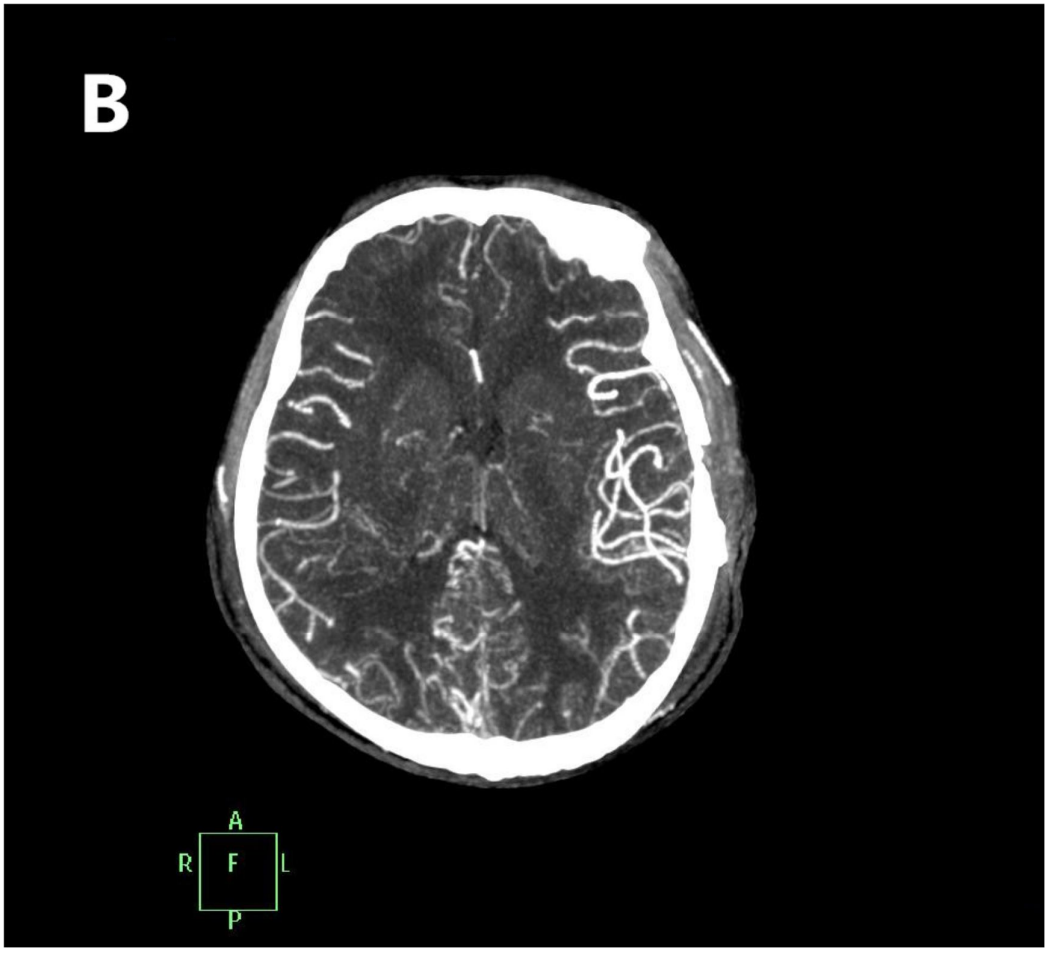
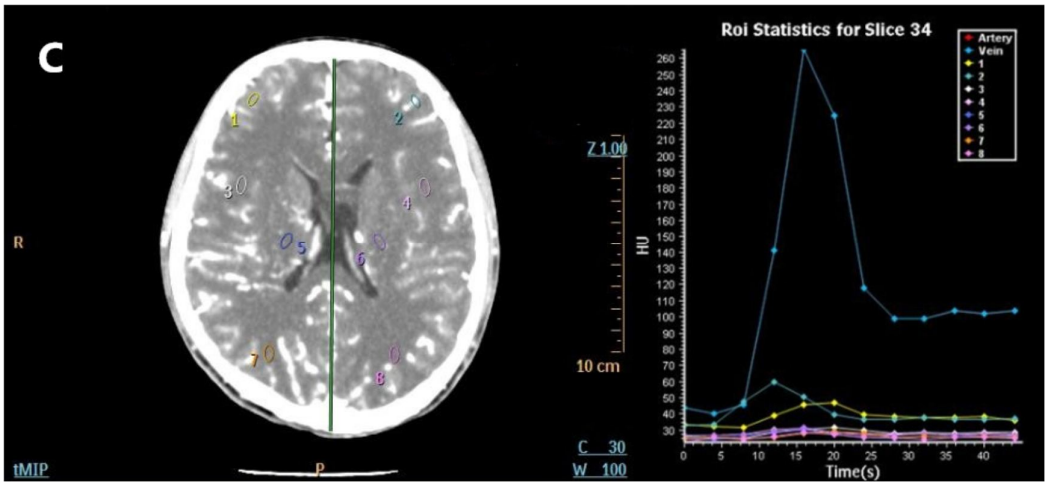
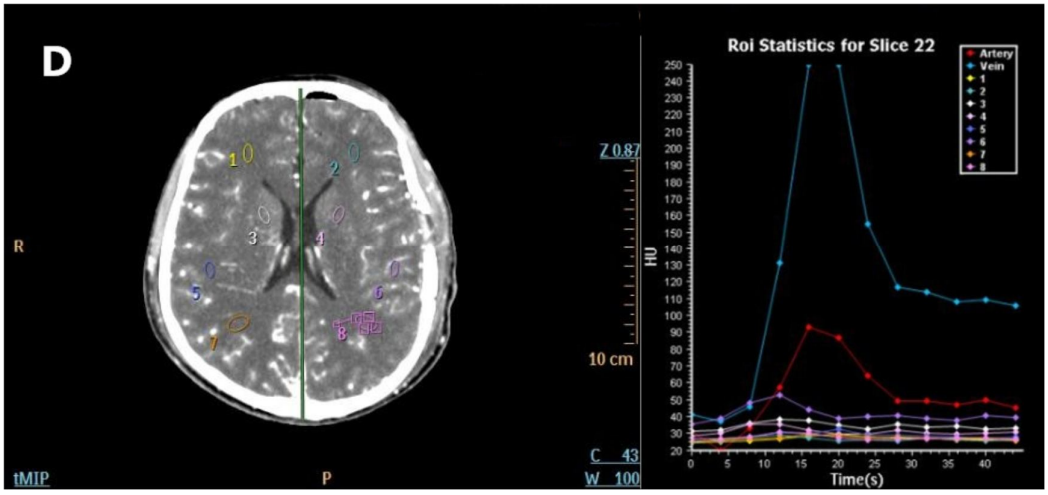


Figure 2. Computed tomography angiography (CTA) and CT perfusion (CTP): (A) pre-operative CTA. (B) two weeks post-operatiive CTA showed newborn compensatory proliferated vasculature. (C) preoperative CTP. (D) two weeks post-operative CTP showed increased cerebral blood flow.









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