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Case Report

Usefulness of combined bypass surgery for moyamoya disease in infants under 1 year of age: A technical case report

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ABSTRACT

Background: Among pediatric cases of moyamoya disease (MMD), cerebral ischemic symptoms often progress and worsen rapidly in infants under one year of age; therefore, it is important to treat them as early as possible. However, direct bypass surgery is often technically difficult for infants due to their small blood vessels. Here, we describe our technique to resolve the technical challenges encountered during superficial temporal artery-to-middle cerebral artery (STA-MCA) bypass surgery in infants aged <1 year with MMD, focusing on specific procedures.

Case Description: We performed bilateral STA-MCA and indirect bypass in a 1-year-old girl with MMD and cerebral infarction. Before treatment, a peripherally inserted central venous catheter (PICC) was placed to avoid ischemic attacks associated with crying, dehydration, and malnutrition. All examinations and procedures that would be stressful to the patient, such as blood examinations, were performed using PICC or under sedation. The STA-MCA diameters were 0.8 and 1.2 mm, respectively. After suturing the planned anastomosis with one stitch using an 11-0 monofilament nylon thread, the thread was lifted upward, and the arterial wall was incised. Anastomosis was performed using an 11-0 monofilament nylon thread with 2–4 stitches on each side. The operation was completed without patency problems. Postoperative blood flow improved, and the patient had a good treatment course.

Conclusion: Direct bypass for MMD patients aged <1 year is technically challenging; however, the vessels can be connected if the procedure is carefully performed with consideration of the characteristics of the infant's vessels.

Keywords: Combined bypass surgery, Infant, Moyamoya disease, Superficial temporal artery-to-middle cerebral artery bypass (STA-MCA bypass)

INTRODUCTION

Moyamoya disease (MMD) is a disease of unknown cause characterized by bilateral stenosis of the internal carotid artery endings. It is a chronic, progressive, and obstructive disease in which abnormal blood channels develop as collateral blood channels.[15] Timely superficial temporal artery-to-middle cerebral artery (STA-MCA) bypass surgery for MMD can improve cerebral hemodynamics, reducing perioperative ischemic complications and eliminating transient ischemic attacks (TIAs) and other ischemic attacks at an early stage.[8,11] However, when revascularization is performed in infants aged <1 year, indirect bypass surgery is often

performed as an alternative due to the procedural difficulties encountered during direct bypass surgery. STA-MCA bypass in infants is a necessary technique but a technically challenging procedure, and there are very few reports in this regard. Therefore, in this report, we describe our technique to resolve the technical challenges encountered during STA-MCA bypass surgery in infants aged <1 year with MMD, focusing on specific procedures.

CASE PRESENTATION

The patient was a 1-year-old girl whose mother and grandmother had a history of MMD. The patient presented with incomplete hemiplegia in the left upper and lower extremities. Magnetic resonance imaging of the head revealed acute cerebral infarction in the white matter of the right cerebral hemisphere [Figure 1a]. Magnetic resonance angiography (MRA) of the head and digital subtraction angiography (DSA) showed unclear delineation beyond the peripheral portions of the bilateral internal carotid arteries, and a diagnosis of MMD was made [Figures 1b and c].[12] The results of external carotid arteriography showed a very thin superficial temporal artery (STA) on both sides [Figure 1d]. Single-photon emission computed tomography with N-isopropyl-p-[123I] iodoamphetamine (^{123}I-IMP) showed reduced blood flow at rest in bilateral anterior and middle cerebral artery (MCA) regions [Figure 1e]. The patient underwent STA-MCA bypass surgery, with indirect bypass on the right side 22 days after the stroke and on the left side 50 days after the stroke. There were no new neurological symptoms after surgery, and the patient was discharged from the hospital after rehabilitation without neurological sequelae.

Three months postoperatively, MRA and DSA showed adequate bilateral blood flow in the bypass area and well-developed collateral blood flow [Figure 1f]. One year postoperatively, the patient was doing well with no apparent neurological symptoms such as TIA.

Approval was obtained from the University's Institutional Review Board before initiation of the treatment procedure.

TECHNICAL NOTES

Preoperative preparation

Before treatment, a peripherally inserted central venous catheter (PICC) was inserted to avoid ischemic attacks

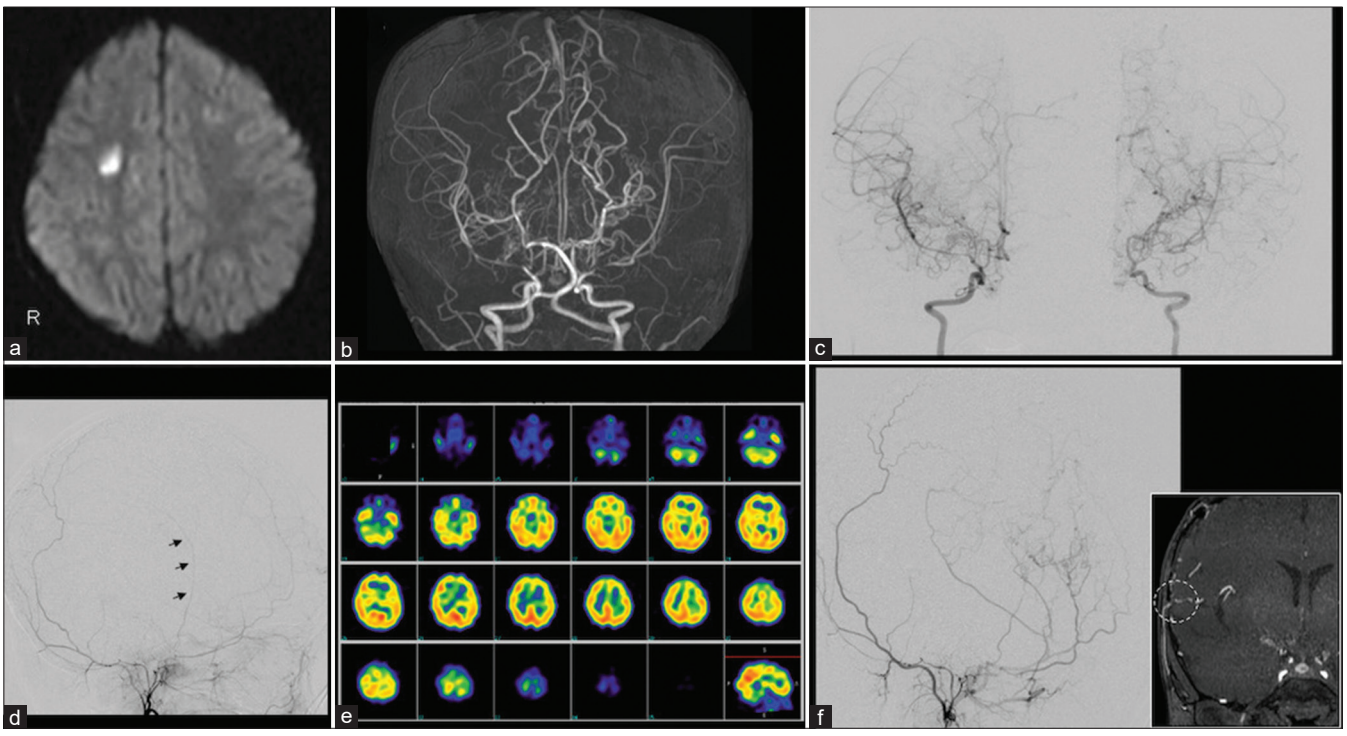
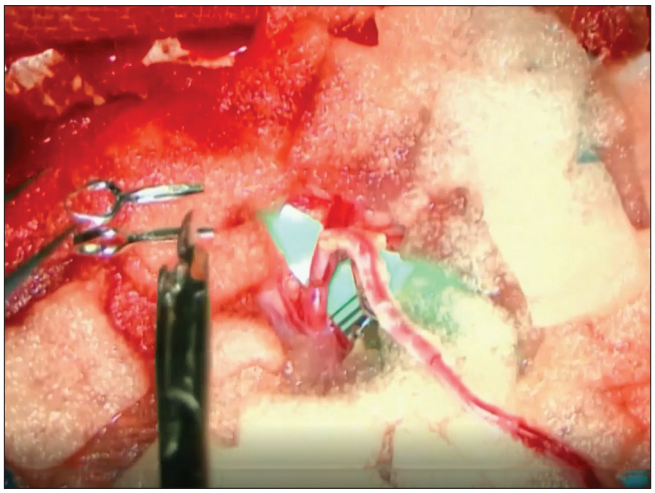


Figure 1: (a) Magnetic resonance imaging of the head showing acute cerebral infarction in the white matter of the right cerebral hemisphere. (b and c) Magnetic resonance angiography (MRA) of the head and digital subtraction angiography (DSA) show unclear delineation beyond the peripheral portions of the bilateral internal carotid arteries. (d) Left external carotid angiography shows that the superficial temporal artery is very thin (arrows). (e) Single-photon emission computed tomography with N-isopropyl-p-[123I] iodoamphetamine showed reduced blood flow at rest in the bilateral anterior and middle cerebral artery regions. (f) MRA and DSA at three months postoperatively showed good blood flow in the bypass area and collateral blood flow development (arrows, dashed circle).

associated with crying, dehydration, and malnutrition. All examinations and procedures that would be stressful to the patient, such as blood examinations, were performed using the PICC or under sedation [Figure 2a]. In addition, patients with MMD take aspirin preoperatively and continue aspirin intraoperatively without discontinuation.



Video 1: Surgery video.

Head fixation

For infants aged <1 year, their skull is thin and should be secured at four points using pediatric pins to reduce the risk of skull penetration [Video 1].

Scalp incision and STA graft harvesting

Using cerebral angiography as a reference, we marked the run of the frontal and parietal branches of the STA on the scalp using palpation and vascular Doppler. To perform a single bypass, a semicircular skin incision was made on a semicircular arc with its base on the auricular side, centered on the parietal branch of the STA, without damaging the main trunk of the frontal branch [Figure 2b]. An incision was made up to the galea, scalp clips were applied, and the skin valve was flipped.

STA grafts were harvested using a microscope. STAs in infants aged <1 year are easily damaged because their diameter is <1 mm and the vessel wall is very thin. However, they are characterized by little flexural tortuosity and vascular branching. Therefore, grafts should be harvested using a combination of low-power coagulation and sharp incision, paying attention to coagulation heat and tear damage during

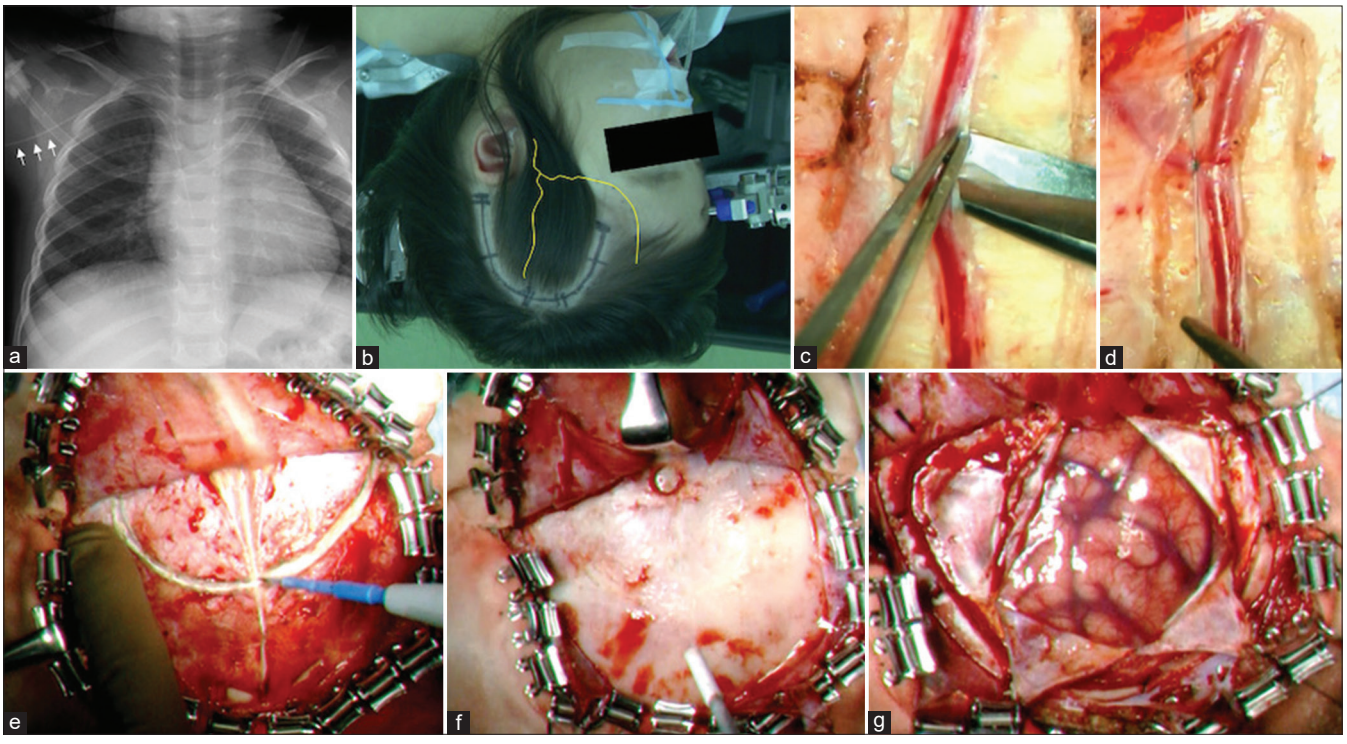


Figure 2: (a) Before treatment, a peripherally inserted central venous catheter was inserted to avoid ischemic attacks due to dehydration, chirping, and malnutrition (arrows). (b) A semicircular arcuate skin incision was made around the parietal branch of the superficial temporal artery (STA) (the yellow line indicates the STA). (c and d) The STA graft was detached by approximately 4 cm, and the thicker branches were ligated using an 8-0 nylon thread, coagulated and cut. (e) The temporal muscle was incised in the middle along the STA graft run. (f) Craniotomy was performed with only one burr hole for the graft tunnel in children aged <1 year. (g) A cross incision was made in the dura mater to avoid obstructing the passage of the STA graft.

dissection [Figure 2c]. STA grafts should be detached by approximately 4 cm, and thick branches should be ligated using an 8-0 nylon thread, coagulated, and cut [Figure 2d]. After the graft was harvested, the proximal end was blocked with a clip, the distal end was cut, and the lumen was flushed with saline solution and heparin to prevent thrombus formation.

Craniotomy and dural incision

The temporal muscle was incised medially along the STA graft run, and a Y-shaped incision was made where the graft passed through the muscle to avoid compression during closure [Figure 2e]. Once the bony surface is exposed, craniotomy is performed with only one burr hole for the graft tunnel to minimize bone defects in children aged <1 year [Figure 2f]. A dural incision should also be made in the midline to avoid interfering with the passage of the STA graft, and a cross incision should be made in the middle to go straight through [Figure 2g]. During a dural incision, care should be taken to avoid damaging the middle meningeal artery.

The end-tidal CO_2 ($EtCO_2$) during surgery should also be noted. Surgery is performed with $EtCO_2$ within normal limits; however, if serious brain swelling is observed when the dura mater is incised, $EtCO_2$ should be lowered.

Recipient preparation

The recipient should be chosen by securing the central or precentral artery of the MCA to distribute bypass blood flow throughout the MCA region as much as possible. After carefully incising the surrounding arachnoid membrane and securing the recipient, a silicone Super Micro Sheet K (Kono Seisakusyo, Tokyo, Japan) was inserted under the recipient to lift the planned anastomosis area to facilitate anastomosis [Figure 3a]. Gelfoam (Pfizer, Tokyo, Japan), cut into small pieces of 10 mm × 10 mm, was placed on the brain surface around the recipient to protect the brain surface and provide space for needle placement [Figure 3b]. Next, the donor was prepared by cutting the peripheral portion of the graft, being aware of its length, for the secured STA graft to reach the recipient with enough room to spare. Approximately 1–2 cm of tissue attached to the STA graft end was removed, and the tip of the graft was trimmed [Figure 3c]. The tip of the STA graft was placed over the recipient, and the intracranial position of the graft was rechecked to ensure that it was anastomosed naturally without twisting. The graft tip was then cut at an angle to allow blood flow from the STA graft to flow naturally. The cut orifices were coated with biological pigment. The diameter of the recipient and STA graft was 1.2 mm and 0.8 mm, respectively [Figures 3d and e].

Patients aged ≤3 Years require caution in anastomosis because the average diameter of the STA graft is 0.8 mm, and the vessel wall is very thin, depending on age. Before

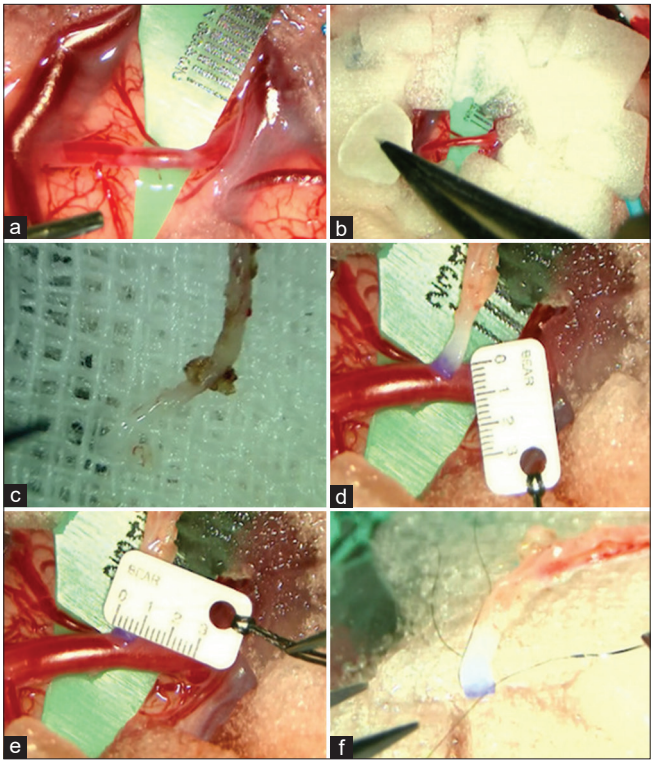


Figure 3: (a) A silicone super micro sheet K was inserted under the recipient. (b) Gelfoam, cut into small pieces of 10 mm × 10 mm, was placed on the brain surface around the recipient. (c) Approximately 1–2 cm of the tissue attached to the superficial temporal artery (STA) graft end was removed, and the graft tip was trimmed. (d and e) The diameter of the recipient and STA graft was 1.2 mm and 0.8 mm, respectively. (f) Before anastomosis, a stay suture needle was threaded through both ends of the STA graft in advance using an 11-0 monofilament nylon thread.

anastomosis, a stay suture needle was threaded through both ends of the STA graft in advance using an 11-0 monofilament nylon thread (BEAR Medic Co., Ibaraki, Japan) [Figure 3f]. This allows the MCA to be blocked as quickly as possible during anastomosis.

Anastomosis

Because patients with MMD take aspirin, hemostasis of the operative field should be thoroughly checked before anastomosis is performed. After marking the anastomotic site of the recipient and the position where the clip was to be applied with biological pigment, the clip was used to block the blood flow [Figures 4a and b]. Infant patients with MMD have fewer pial arteries branching off the recipient due to their anatomy; however, during coagulation and cutting, bipolar should be used at a very low power to avoid heat coagulation of the recipient.[2]

After suturing the planned anastomosis with one stitch using an 11-0 monofilament nylon thread, the thread was

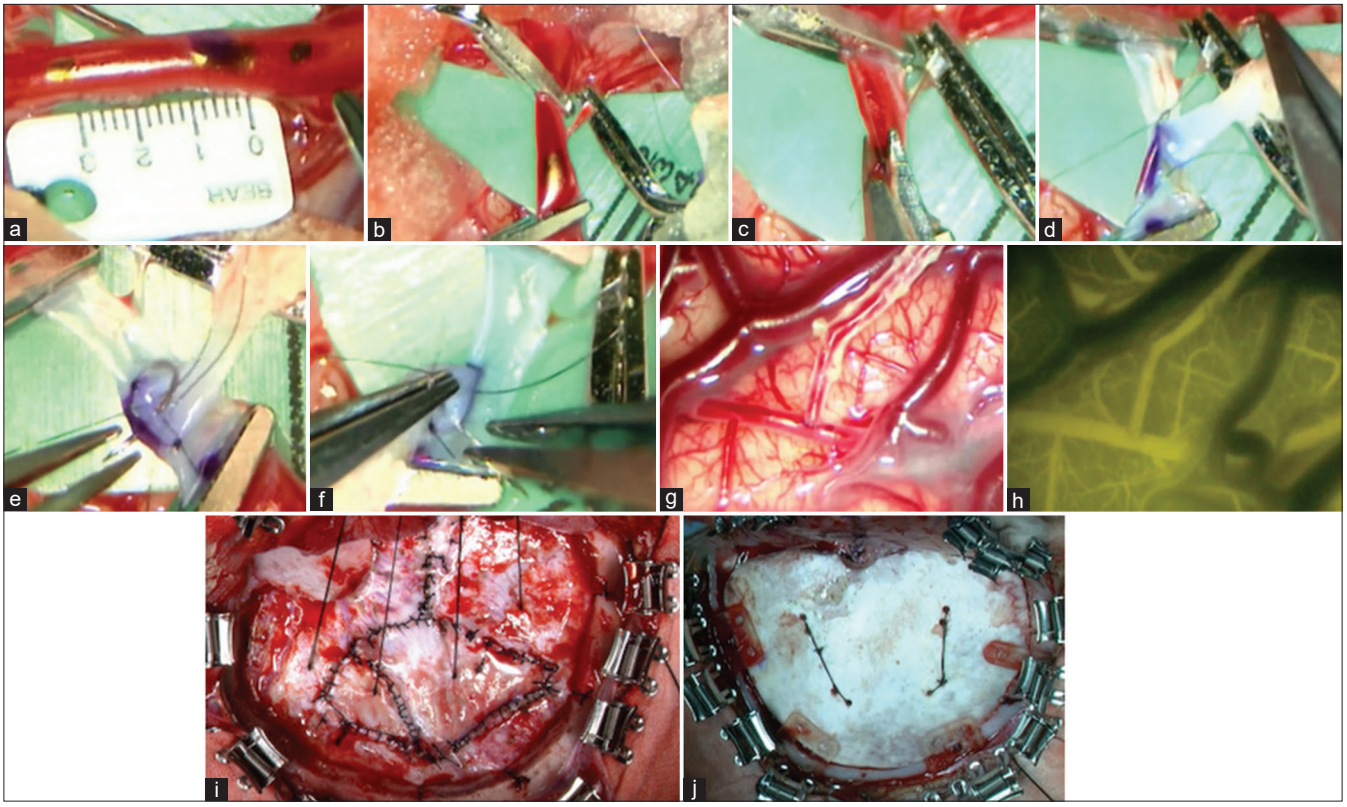


Figure 4: (a and b) After marking the anastomotic site of the recipient and the position where the clip is to be applied with biological pigment, the clip was used to block blood flow. (c) After suturing the planned anastomosis with one stitch using an 11-0 monofilament nylon thread, the thread was lifted upward, and the arterial wall was incised. (d) A stay suture was first performed at both ends. (e and f) Anastomosis was performed with interrupted sutures. Anastomosis was performed with 2–4 stitches on each side. (g and h) After anastomosis, micro-Doppler and indocyanine green were used to confirm adequate blood flow in the bypass area. (i) The dura mater was folded back to the brain's surface for encephalo-duro-synangiosis. The dural defect was replaced by harvesting of the temporal fascia. (j) The bone flap was fixed using an absorbable osteosynthesis plate in the infant.

lifted upward, and the arterial wall was incised [Figure 4c]. The lumen was rinsed with heparinized saline, and after confirming that there was no leakage of arterial blood, a stay suture was first performed at both ends [Figure 4d]. Anastomosis was then performed on one side with an interrupted suture. After threading the needle at all points, the back was checked for backstitching without ligature. If ligation is performed first, there is a possibility of injuring blood vessels when cutting the ligatured thread if a backstitch is used. Anastomosis was performed with 2–4 stitches on each side [Figures 4e and f]. The clip was then removed, and the blood flow resumed. If there is a blood leak from the anastomosis, additional sutures or attaching a small piece of harvested fascia will almost always stop the bleeding.

From post bypass to closed head

After anastomosis, micro-Doppler and indocyanine green were used to confirm adequate blood flow in the bypass area [Figure 4g and h]. If a thrombus is found in a bypass vessel, mechanical disruption with a settler or intravenous administration of heparin, argatroban, or ozagrel sodium should be performed.

The dura mater is folded back to the brain's surface for encephalo-duro-synangiosis for indirect bypass. The dural defect was replaced by harvesting of the temporal fascia [Figure 4i]. The bone flap was fixed using an absorbable osteosynthesis plate in infants [Figure 4j].

DISCUSSION

We presented a detailed description of STA-MCA bypass surgery in an infant aged <1 year with MMD, focusing on the technical aspects of the procedure and specific surgical procedures. Various revascularization techniques are used for MMD and can be broadly classified into direct and indirect revascularization. Controlled clinical trials have not reported significant differences in outcomes among surgical procedures for pediatric MMD; however, the frequency of ischemic stroke after indirect revascularization has been reported to be 7.7–13.7%.[10,13] Conversely, the frequency of ischemic stroke

in the perioperative period of combined direct and indirect revascularization surgery in Japan has been reported to be 0–5.5%.[9] Therefore, despite regional and ethnic differences, combined direct and indirect revascularization surgery as early as possible is considered necessary for pediatric MMD. However, direct revascularization for infants aged <1 year is generally performed using indirect revascularization because the vessels are very thin, and the procedure is very difficult. Indirect bypass is less difficult to perform, and collateral vessels develop better in children. However, the disadvantage is that it takes a few months after the procedure for the collateral vessels to grow, and a small number of cerebral infarctions have been reported before the indirect bypass is functional.[5,10,13] Therefore, direct revascularization is needed to improve cerebral hemodynamics, even earlier in infants aged <1 year.[4,6,7,10,13]

When performing direct revascularization for MMD in infants aged <1 year, a PICC should be inserted before treatment, and tests and procedures that may be stressful to the patient should be performed using PICC or under sedation, as appropriate to avoid ischemic attacks associated with crying, dehydration, and malnutrition. Preoperative medical therapy includes aspirin at a dose of body weight × 2 mg/day and continued oral administration during and after surgery.[14,16] The first technical note in surgery is that infants aged <1 year have anatomically very thin and soft STA grafts and recipient vessel walls compared with adults and schoolchildren.[1] Therefore, when the clip is applied to both sides of the recipient, the load crushes the recipient and flattens it, resulting in a high risk of backstitching. To solve this problem, we believe that this method, in which all threads are first threaded on one side and then ligated after checking the opposite side for backstitching, is more effective in infants. Furthermore, when penetrating the vessel wall with an 11-0 monofilament nylon thread, the most important thing to consider is that infants are easily dissected or torn, which can injure blood vessels.[1] Therefore, when penetrating the needle, it is necessary to always move the hand with an awareness of the force vector perpendicular to the vessel wall. When sewing on the opposite side, it is usually better to operate the needle with the non-dominant hand because the direction of the hand is usually reversed, making suturing difficult. Arteriectomy in the recipient must also be performed with care; our recommended method involves the use of an 11-0 monofilament nylon thread that is lifted upward after one suture at the planned anastomosis site. This is because the vessels in infants with MMD are so thin and soft that any attempt to incise them without lifting the vessel wall with a thread can easily collapse the vessel and damage the contralateral vessel wall. Finally, regarding the ligation method, the interrupted suture is recommended over a continuous suture.[3] This is because the thread is so thin that in the case of continuous sutures, the force of closure is weak, and the thread is not very durable. In addition, if one thread breaks or loosens, it will loosen in all areas, making recovery difficult for infants with thin vessel walls.

CONCLUSION

Direct bypass for MMD patients aged <1 year is technically challenging; however, the vessels can be connected if the procedure is carefully performed, considering the characteristics of the infant's vessels.

Acknowledgments

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Ethical approval

The research/study was approved by the Institutional Review Board at the Ethical Committee for Epidemiology of Hiroshima University, number E2022-0016, dated February 16, 2023.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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