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CLINICAL REPORT

Deep hypothermic circulatory arrest for hemiarch replacement in a pediatric patient with moyamoya disease

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Abstract Moyamoya disease is a chronic cerebrovascular occlusive disease, occurring predominantly in young populations, that causes cerebral ischemia and hemorrhage. Patients with moyamoya disease are at high risk of neurological complications during cardiac surgery because of perioperative hemodynamic changes. However, there is no established evidence on temperature management during cardiopulmonary bypass. Previous reports described normothermia or mild to moderate hypothermia during cardiopulmonary bypass in patients with moyamoya disease; however, surgical conditions, such as not having enough space to clamp the aorta or a clean surgical field, sometimes force us to use deep hypothermic circuratory arrest. We report a successful case of a pediatric patient with moyamoya disease who underwent deep hypothermic circulatory arrest (18 °C) for hemiarch replacement without neurological complications. Deep hypothermia may be an alternative technique for achieving cerebral protection in the context of moyamoya disease.

Keywords Moyamoya disease · Deep hypothermic circulatory arrest · Cardiopulmonary bypass

Introduction

Moyamoya disease (MMD) is a chronic progressive cerebrovascular disease involving occlusion of the internal carotid arteries and development of collateral networks [1]. MMD presents predominantly in young populations in Japan and Asian countries, and causes not only cerebral ischemia but also intracranial hemorrhage because the collateral vessels are fragile and small compared with normal vessels. These patients are therefore at high risk of neurological complications, especially during cardiac surgery as a result of perioperative hemodynamic changes [2]. However, reports regarding cardiac surgery in MMD cases are limited, and there is no established evidence on temperature management during cardiopulmonary bypass (CPB). Previous reports stated that patients underwent normothermic CPB (35–36 °C) [3, 4] or mild to moderately hypothermic CPB (29–34 °C) [2, 5, 6] without neurological sequelae. Although surgical conditions such as not having enough space to clamp the aorta or sufficient clean surgical field sometimes force us to use deep hypothermic circuratory arrest (DHCA), there are no data on DHCA in MMD. DHCA has the advantage of cerebral protection and a clean surgical field, but cerebral autoregulation is impaired during both cooling and warming phases [7], suggesting there may be an additional higher risk of stroke in MMD. We report a successful case of a pediatric MMD patient who underwent hemiarch replacement during DHCA without neurological complications.

Case report

A 9-year-old girl (height 122 cm, body weight 21 kg) was referred to our institution for an ascending aortic aneurysm. Previous genetic screening revealed a mutation in the ACTA2 gene, which codes for smooth muscle α-2 actin in vascular smooth muscle cells. ACTA2 mutations are associated with increased risk of thoracic aortic aneurysms,

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MMD, and coronary artery disease [8]. The patient had a history of patent ductus arteriosus ligation at 3 months old and a patch repair for aneurysmal dilatation of ligated ductus arteriosus at 1 year old. Computed tomography showed marked dilation of the aortic root and the ascending aorta (maximal diameter, 44 mm), as shown in Fig. 1, and transthoracic echocardiography showed mild aortic regurgitation caused by annuloaortic ectasia. Cerebral magnetic resonance angiography revealed occlusion of the right internal carotid artery and distal small arteries (Fig. 2). In addition, single photon emission computed tomography with acetazolamide stimulation demonstrated reduced cerebral blood flow in the bilateral temporal and occipital lobes at rest and decreased cerebrovascular reactivity globally (especially in the bilateral frontal lobes), indicating cerebral hemodynamic impairment comparable to Powers' stage 2 [9], which represents an inadequate blood supply relative to metabolic demand (misery perfusion [10]). The patient was diagnosed with MMD despite not presenting with neurological symptoms. Pediatric cardiac surgeons, vascular surgeons, and anesthesiologists held a joint conference to determine the surgical strategy. We decided to perform ascending aortic replacement and plication of the sinotubular junction under moderate hypothermic CPB [6] to minimize the risk of cerebral injury.

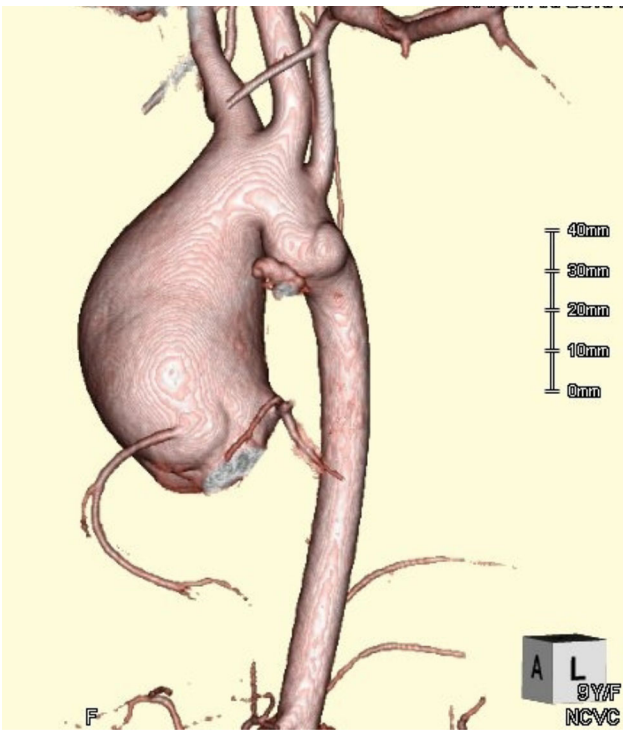

Fig. 1 Computed tomography image shows dilation of aortic root and ascending aorta

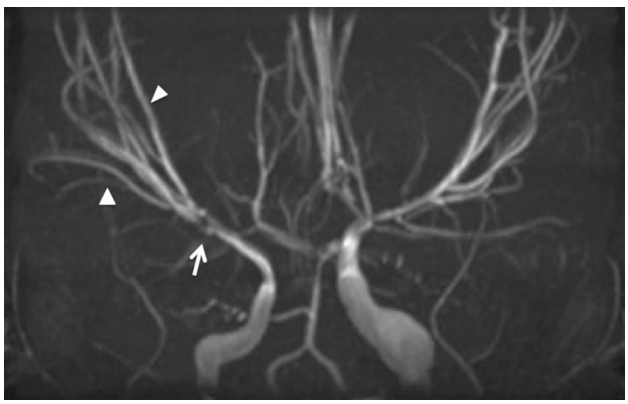

Fig. 2 Magnetic resonance angiography image shows occlusion of right internal carotid artery (*arrow*) and distal small arteries (*arrowheads*)

General anesthesia was slowly induced with sevoflurane 5 % and nitrous oxide 60 %, and maintained with propofol 1–7 mg/kg/h, remifentanil 0.1–0.5 μg/kg/min, and fentanyl (total, 800 μg). We did not use sevoflurane for maintenance of anesthesia to avoid the risk of the steal phenomenon from cerebral vasodilation by sevoflurane [11], because this phenomenon could deteriorate cerebral ischemia as the result of reduced cerebrovascular reactivity caused by MMD. Tranexamic acid was continuously infused at 400 mg/h during the surgery. We also planned to control the partial pressure of carbon dioxide in arterial blood ($PaCO_2$) at normocapnea and to adopt α-stat management during CPB. $PaCO_2$ was 40–43 mmHg before and after CPB but about 50 mmHg during CPB.

Monitoring included electrocardiogram, right radial arterial pressure, capnography, pulse oximetry, central venous pressure, and body temperature (tympanic, bladder, and palmar temperatures). We also attached a bispectral index (BIS) electrode (QE-910P; Aspect Medical Systems/Nihon Kohden, Tokyo, Japan) as well as two sensors for near-infrared spectroscopy (NIRS) (NIRO200NX; Hamamatsu Photonics, Hamamatsu, Japan) at the right and left forehead to monitor regional cerebral oxygen saturation (rSO_2). CPB was established uneventfully, and the patient was cooled to a tympanic temperature of 30 °C. Perfusion pressure was 40–45 mmHg at the start of CPB and increased to 55–65 mmHg with continuous infusion of norepinephrine.

Ascending aortic replacement and plication of the sinotubular junction were performed as scheduled, but bleeding from the distal anastomosis between the ascending aorta and the artificial graft could not be controlled because of vascular fragility. We therefore immediately decided to remove the fragile component and to change the surgical procedure to hemiarch replacement. Temperature management was converted from moderate hypothermia to DHCA because it was a challenge to clamp the aortic arch and to obtain a clear surgical field. The patient was rapidly

cooled to 18 °C. We also cooled the patient's head with an ice pack and started to administer a loading dose of 500 mg for 30 min followed by continuous infusion of an additional 500 mg until the end of the surgery to protect the brain [12]. Hemiarch replacement was performed in DHCA with retrograde cerebral perfusion (RCP) [13]. After separation from prolonged CPB, massive transfusion was required to control bleeding (16 units of red blood cells, 24 units of fresh frozen plasma, 40 units of platelets, and 300 ml of cryoprecipitate). The BIS values were maintained at 50–60 before CPB and decreased to 20–30 after institution of CPB. After conversion to DHCA, BIS values declined to around 0, then gradually increased to 20–40 with rewarming. NIRS showed a discrepancy between right and left rSO_2 during CPB (including DHCA), but the bilateral rSO_2 value remained above 60 % throughout almost the entire operation (Fig. 3). The operative time was 14 h 48 min, total CPB duration was 9 h 6 min, and DHCA duration was 45 min. The anesthetic record was shown in Fig. 4. After the operation, the patient was transferred to the intensive care unit. She was extubated on postoperative day 2 and discharged from the intensive care unit on postoperative day 4. She had no neurological complications postoperatively.

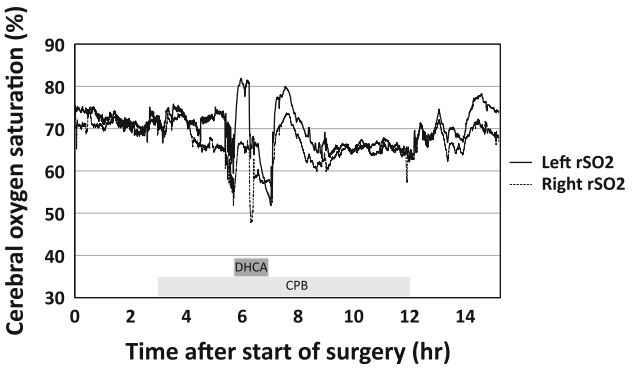

Fig. 3 Intraoperative regional cerebral oxygen saturation measured by near-infrared spectroscopy. *DHCA* deep hypothermic circulatory arrest, *CPB* cardiopulmonary bypass, rSO_2 regional cerebral oxygen saturation

Discussion

This patient had a history of aneurysmal dilatation of ligated ductus arteriosus and also suffered concurrently from MMD and an ascending aortic aneurysm. These disorders were potentially caused by a mutation in the ACTA2 gene. ACTA2 mutations are associated with dysfunction of vascular smooth muscle, also described in other connective tissue disorders such as Ehlers–Danlos syndrome [14], and might have contributed to her vascular fragility and easy bleeding. MMD is a cerebrovascular occlusive disease and causes both cerebral ischemia and hemorrhage [1]. CPB is associated with a high risk of these neurological complications because many perioperative factors influence cerebral hemodynamics, such as decreased hematocrit from

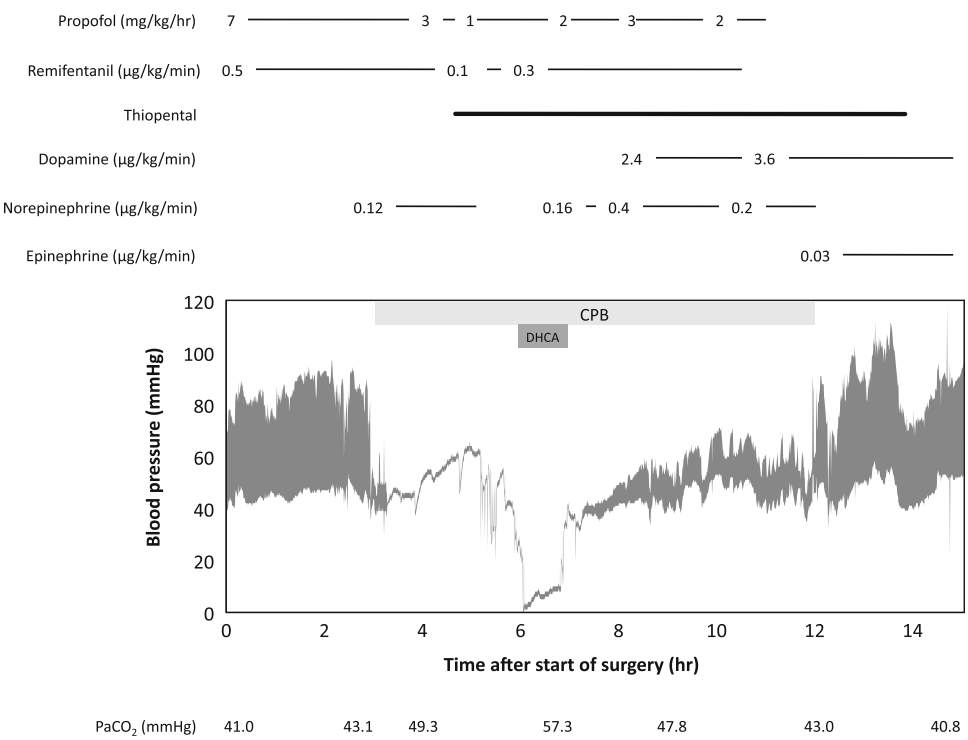

Fig. 4 Anesthetic record. *DHCA* deep hypothermic circulatory arrest, *CPB* cardiopulmonary bypass, rSO_2 regional cerebral oxygen saturation, $PaCO_2$ partial pressure of carbon dioxide in arterial blood

dilution, nonpulsatile CPB flow and changes in perfusion pressure, and CPB-induced dysfunction of the blood–brain barrier [15]. In regard to the DHCA procedure, vasoconstriction caused by hypothermia may be harmful during the cooling phase as decreases in cerebral blood flow may lead to cerebral ischemia in MMD patients. Cook et al. [16] demonstrated that the cerebral metabolic ratio of oxygen decreased significantly more than did the cerebral blood flow, resulting in cerebral protection. However, there is no report of MMD patients undergoing DHCA, and temperature management in CPB still remained to be resolved. Some authors have stated that mild to moderately hypothermic CPB is favorable in MMD because hypothermia can decrease cerebral oxygen demand and provide cerebral protection [3, 4]. Others have recommended normothermic CPB because even mild hypothermia may cause constriction of collateral vessels, leading to risk of cerebral ischemia in MMD [2, 5, 6]. None of these studies reported any neurological complications, indicating a potential advantage of cerebral protection over vasoconstriction during hypothermia. Further, we used the RCP technique during DHCA. RCP can prolong the safe time of cerebral protection compared with DHCA only [13]. Antegrade selective cerebral perfusion is also an effective method for cerebral protection [13]. However, hypoperfusion and hyperperfusion during antegrade selective cerebral perfusion may cause cerebral ischemia and hemorrhage, respectively, in fragile cerebral vessels of MMD, and the optimal flow of selective cerebral perfusion is unclear in patients with MMD. In addition, the technical simplicity of RCP is suitable for unexpected or emergent introduction of DHCA as shown in this case. For pharmacological cerebral protection, we administered thiopental immediately after the decision to convert to DHCA. Thiopental decreases cerebral metabolism and oxygen demand and provides cerebral protection [17]. NIRS showed a discrepancy between right and left rSO_2 during CPB (including DHCA); this may be caused by occlusion of the right internal carotid artery, but the precise mechanism remains unclear.

In this case DHCA was introduced unexpectedly, and its management therefore differed from that in previous reports, for instance, high $PaCO_2$, prolonged CPB time, and massive transfusion. Both hypercapnea and hypocapnea are risk factors for cerebral ischemia, and normocapnea is recommended in MMD patients [18, 19]. We could control $PaCO_2$ at normocapnea before and after CPB. However, the patient was exposed to an elevated $PaCO_2$ (about 50 mmHg) despite α-stat management during CPB, partly because carbon dioxide insufflated in the pericardial cavity might be suctioned and mixed with the blood in the CPB reservoir, which might have an effect on cerebral blood flow. Prolonged CPB time and massive transfusion could cause unstable cerebral perfusion. However, the strategies used to protect the brain, as described here, might have contributed to postoperative recovery without neurological complications.

In conclusion, this is a report of a pediatric MMD patient undergoing DHCA for hemiarch replacement. With careful cerebral monitoring using BIS and NIRS, DHCA may be an alternative technique for cerebral protection of MMD patients in cases in which the DHCA technique is required for major vascular surgery.

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