

ANAESTHETIC MANAGEMENT FOR INGUINAL HERNIA REPAIR IN A CHILD WITH MOYAMOYA DISEASE

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Abstract:

Moyamoya is probably an inherited vaso-occlusive disease, first described in Japan and now reported from all over the world. We describe a case of a 9 year old boy, receiving general anaesthesia for a scheduled inguinal hernia repair. At 7.5 years, the boy was successfully remedied for symptomatic moyamoya disease by bilateral encephalo – duro – arterio – synangiosis (EDAS).

Key Words: CBF; ICP; LMA; moyamoya disease; sevoflurane

Introduction

Moyamoya disease or syndrome is an unusual form of chronic, progressive occlusive cerebrovascular disorder, first described in Japan and now reported from all over the world that usually presents as transient, recurrent hemispheric or ischaemic attacks – strokes, mostly in children, during the first decade of life¹. Distal internal carotid, basilar arteries and their vicinity, as well as proximal portions of the anterior and middle cerebral arteries gradually narrow and or close, leading to proliferation of penetrating arteries, primarily at the base of brain².

These abnormal dilated vessels (moyamoya vessels) are a source of a hazy network of collateral flow and circulation to the base of the ischaemic brain, distal to the stenotic process, and produce a net – like image on cerebral angiograms³. Thus, moyamoya syndrome, occurring as a result of a sudden cerebrovascular insufficiency, represents an occlusive process, usually bilateral, at the termination of the internal carotid artery and the circulus arteriosus cerebri (circle of Willis). In this disease, a web of small collateral blood vessels in the post – stenotic area is seen in carotid angiography⁴.

In the sixties, Suzuki and Takaku named this disease moyamoya, which in Japanese means “something hazy, like a wavering puff of cigarette smoke, drifting in the air”. This term was used to describe the abnormal vasculature at the base of brain, as it was briefly analyzed above⁵.

Given the chronic nature of cerebral ischaemia and the debilitating course of moyamoya disease, various surgical procedures have been proposed to augment collateral cerebral blood flow (CBF)^{6,7,8,9}.

CBF in children with moyamoya syndrome is severely compromised¹. This may account for the cerebral ischaemia, mostly due to asymmetrical involvement of bilateral internal carotid arteries (ICAs) and posterior cerebral artery systems (PCA) and to the predominant implication of the ipsilateral ICAs and PCA that primarily occurs in children afflicted with this disease¹⁰. This lack of blood may cause hemiplegia, paralysis of the feet, legs or the upper extremities. Headaches, various vision problems, mental retardation and psychiatric problems may occur. Adults sticken with moyamoya syndrome primarily develop intraventricular haemorrhages¹.

Therefore, as haemodynamic changes are often during surgery and as ventilatory alterations and adjustments are subject to the anaesthesiologist’s hands, intensive anaesthetic management during surgical operations in these patients, or even during interventional procedures, is needed to maintain CBF and minimize the risk of neurological complications. such as new focal neurological deficits.

alterations in mental status, perioperative strokes, cerebral infarctions or intracranial haemorrhage^{11,12}. Hypertension easily causes intracranial haemorrhage, and hypotension or hyperventilation easily brings about cerebral ischaemia¹³.

We report a case of general anaesthesia with sevoflurane and the application of laryngeal mask (LMA) for scheduled inguinal hernia repair, in a child successfully operated for moyamoya disease in the past, and we describe the whole anaesthetic management in this operation of short duration.

Case report

A 9 – year – old boy, 35 kg weight, 145 cm height, presented to our hospital for a scheduled operation of inguinal hernia repair. In the past, the boy suffered from symptomatic Moyamoya disease and right persistent primitive trigeminal artery, which causes multiple cerebral infarctions. To prevent the progression of the disease he was admitted in Tokyo Medical and Dental University, Japan, when he was 7.5 years old, for surgical treatment. Bilateral encephalo – duro – arterio – synangiosis – EDAS (indirect anastomosis) was performed under general anaesthesia, with uneventful postsurgical course.

At that period of time the first symptom was generalized tonic – clonic seizures twice, at the age of seven years old. Despite antiepileptic medication, the child suffered from focal seizures, migraine and two episodes of post – seizure paresis. One and a half year after surgical treatment he was free of motor symptoms, transient ischaemic episodes and epileptic seizures, while his IQ was normal. His present medication was 50 mg of phenobarbital, per os, twice per day. Thus, postoperatively, the child's neurologic symptoms were resolved and possible seizure activity was controlled by oral anticonvulsant medication.

The child was admitted to our hospital for a scheduled inguinal hernia repair. Preanaesthetic assessment was performed, as well as discussion with surgeons and neuropaediatricians concerning the whole procedure.

The child came to the operating room calm, after being premedicated with rectally administered midazolam (0.1 mg/kg), 30 minutes prior to anaesthesia induction. A 20 G venous cannula was inserted, after Emla cream 2.5% presetting. The i.v. fluid administration was NS 0.9 % (10 ml/kg) intraoperatively. General anaesthesia was induced with i.v. administration of atropine 0.02 mg/kg, thiopental 5 mg/kg, fentanyl 3 mcg/kg and vecuronium 0.1 mg/kg. The patient's level of sedation and the tolerance of anaesthesia face mask were satisfactory. Preoxygenation with 100% O₂ through the face mask and anaesthesia induction were performed smoothly. An

LMA No 3 was easily inserted and ventilation was controlled mechanically. Anaesthesia was maintained with sevoflurane at an inspired concentration of approximately 2% with mixture of N₂O/O₂ 50:50 (6 lit/min). Prior to surgical procedure, paracetamol suppository (30 mg/kg) was administered rectally.

The intraoperative monitoring included non-invasive measurement of arterial blood pressure (NIBP) every 5 min, as well as electrocardiogram (ECG), heart rate (HR), oxygen saturation (SpO₂) and end - tidal carbon dioxide (EtCO₂), being recorded continuously. The depth of anaesthesia was controlled to maintain normotension appropriate for age. Ventilation was adjusted to maintain normocarbida by closely measuring EtCO₂. Intraoperatively, we maintained EtCO₂ around 34 - 36 mm Hg, NIBP between 110 - 125 mm Hg and 60 - 75 mm Hg for systolic and diastolic blood pressure respectively, HR around 85 bpm and SpO₂ 100% approximately. The duration of the operation was 20 min. At the end of the procedure the LMA was removed uneventfully, without using neuromuscular blocking agents' reversals.

The child was transported to the Post Anaesthesia Care Unit (PACU) and remained there for one and a half hour under observation and routine monitoring. The main considerations were good oxygenation, achieved with the use of a simple mask (O₂ 12 lit/min) and postoperative analgesia. For postoperative analgesia paracetamol suppositories were used. As soon as the child was fully awake, without any signs or symptoms of neurological deterioration and without feeling any pain, he was admitted to the ward. The whole surgical course was uneventful with no postoperative complications and the child was discharged from hospital on the 2nd postoperative day. In a 3 - months period of follow - up no neurological findings suggestive of derangement were referred.

Discussion

Moyamoya disease or syndrome is a disease characterized by the spontaneous occlusion of one or usually both internal carotid arteries in the intracranial region resulting in the formation of a fine network of neovascularity (blood vessel growth) at the base of the brain. This syndrome was initially recognized as an angiographic pattern. Thus, magnetic resonance angiography usually reveals abnormal vascular moyamoya networks observed in the vicinity of the previously mentioned areas in the arterial phase, and these abnormal findings are present bilaterally. The large and proliferating irregular vessels and transdiploic collaterals of the external carotid artery that supplies the ischaemic brain essentially cause the moyamoya network, which can be seen at different sites^{1,14,15}.

The syndrome has a higher incidence in Japan, but may also be found in all nationalities, especially in China and Korea^{16,17}. This disease is an intriguing one and little is known about its pathogenesis. Both congenital and acquired categories have been proposed. Hereditary factors may also be important^{1,13,18}. The acquired form of the disease could be associated with neurofibromatosis, tuberous sclerosis, sickle cell anaemia, meningitis, retinitis pigmentosa, fibromascular dysplasia, arteriosclerosis, Down's syndrome, Fanconi's anaemia and radiation therapy to the skull base in children¹.

Moyamoya disease has two age peaks: juvenile (< 10 years) and adult (3rd decade). In children the symptoms are: a) ischaemic, consisting of recurrent, sometimes alternating, episodes of focal cerebral deficit, especially weakness, but also speech and sensory symptoms (these are the most typical symptoms). b) seizures c) involuntary movement disorders d) intracranial haemorrhage. In adults the main sign and symptoms are: a) intracranial haemorrhage, the most common presenting event and b) ischaemic symptoms, such as those in children¹.

CBF is decreased in patients with moyamoya syndrome than in healthy individuals and the morbidity of such patients is directly related to cerebral blood flow¹. In studies using Xenon – 133 inhalation and positron emission tomography, Kuwabara et al. reported that the regional oxygen extraction ratio was greater in children with moyamoya syndrome. The cerebral blood flow was decreased mostly in the frontal region. After hyperventilation the blood flow was reduced in all regions, while cerebrovascular response to hypercapnia was shown to be impaired^{1,19}.

The relationship between partial pressure of arterial carbon dioxide (PaCO₂) and CBF is well documented, but the effects of ventilation on CBF in the presence of intracranial disease are complex. The resulting phenomena, one of which is called intracerebral steal phenomenon, and the other inverse steal phenomenon or Robin Hood syndrome, have been described^{13,20}. In addition, some reports show that in patients with moyamoya syndrome, due to their precarious cerebral circulation, hypercapnia may be detrimental to the cortical circulation and that normocapnia during anaesthesia is more preferable, since it successfully led to the absence of any new neurological deficits^{21,22}.

On the other hand, hypocapnia could also be harmful in such patients, since in moyamoya patients with normal cardiopulmonary function, the significant factor causing brain ischaemia seems to be hyperventilation²³. Carbon dioxide is a potent modulator of cerebrovascular tone and hypocarbia normally produces a reduction in CBF. Tagawa et al. observed a marked reduction in regional CBF when

PaCO₂ decreased below 29 mm Hg. During hyperventilation, hypocarbia caused constriction of the normal cerebral blood vessels. This resulted in decreased regional CBF and regional cerebral hypoxia in the diseased hemisphere due to "steal" from the moyamoya collateral vessels to the dilated cortical vessels after the termination of hyperventilation²⁴.

Although, all the above abnormal alterations seem to be reversed after reperfusion surgery, given the potentially harmful effects of hypo - and hypercapnia in children with moyamoya syndrome, even surgically corrected, we carefully controlled ventilation to achieve a normocarbia state, by closely monitoring EtCO₂, in order to avoid detrimental adverse events in the perioperative period, for the above - named reasons.

One could disagree with the use of premedication in the referred patient, since midazolam could decrease respiratory rate and could lead to hypoventilation and hypercapnia. The provided dose was safe, ensuring an adequate level of sedation prior to anaesthesia induction, without resulting in haemodynamic or respiratory depression and has already been used for preoperative sedation in childhood moyamoya disease. Rectally administered midazolam, as premedication, in 26 cases of moyamoya syndrome resulted in satisfactory sedation level, tolerance of the anaesthesia mask, smooth anaesthesia induction and excellent anterograde amnesia, without any remarkable complications, including cardiovascular and pulmonary deterioration²⁵. Although further investigation of midazolam appropriate dose for premedication is needed, up to 1mg/kg rectally administered seems to be useful in children with moyamoya disease, in which crying and anxiety could trigger or exacerbate their potential neurological findings.

From the anaesthesiologist's point of view, in patients and especially children with moyamoya disease, the main factors of interest should be avoidance of cerebral ischaemia and of intracranial haemorrhage. Thus, the key points of anaesthetic management are avoidance of hyper or hypoventilation (as explained above), maintenance of adequate CBF, normalization of ICP, avoidance of both cerebral vasoconstriction and vasodilation and minimization of arterial blood pressure alterations²⁶.

Ogawa et al. reported that the autoregulatory response to hypotension is substantially diminished in children because they have significant mismatching of CBF and cerebral metabolic rate of O₂ (CMRO₂) and that they are also prone to developing neurologic deficits during hypotensive episodes²⁷. The optimal management of patients with cerebrovascular disease is dependent on balancing CMRO₂ and CBF. Therefore, such a level of anaesthesia should be provided that would decrease the relatively high CMRO₂ in children, while

maintaining adequate CBF. Factors that increase $CMRO_2$, such as painful stimuli (laryngoscopy, tracheal intubation and surgical incision), should be minimized by the use of adequate levels of anaesthesia and analgesia. It has already been proven that laryngoscopy and intubation violate the patients' protective airway reflexes and predictably lead to hypertension, tachycardia and increased ICP²⁸. This is the reason we avoided tracheal intubation and preferred the LMA use. However, it could be mentioned that the patient probably had endotracheal intubation during the first surgical treatment for the disease 1.5 years ago and that it was probably successful. Therefore, one could also wonder how it was proved that LMA was better than other airway. There is no way for sure for proving it, but it is already accepted that the painful stimuli and the stress response being produced after endotracheal intubation are much more greater in comparison with LMA use²⁸. Thus, even in operations of short duration, as this described, and in patients with moyamoya disease, the anaesthesiologist should always be careful with parameters that could potentially compromise cerebrovascular balance.

We also reassured adequate intraoperative and postoperative analgesia by using paracetamol suppositories, which up to date are not susceptible of interfering with cerebrovascular dynamics²⁹. To ensure adequate CBF, we avoided intraoperative hypotension, hypo or hyperventilation and utilized a balanced anaesthetic technique.

We chose thiopental as an induction agent, due to its neuroprotective properties. Thiopental, apart from its hypnotic and anticonvulsant effects, results in reduction of $CMRO_2$ and CBF in a dose – dependent manner. $CMRO_2$ reduction happens uniformly throughout the brain. $CMRO_2$ is depressed slightly more than CBF, such that metabolic supply exceeds metabolic demand. Because barbiturate – induced cerebral vasoconstriction occurs only in normal areas, these agents tend to redistribute blood from normal to ischaemic brain areas (reverse steal phenomenon), leading to unaffected cerebral vasculature of ischaemic brain²⁰. Thus, the use of thiopental in moyamoya syndrome seems to be useful, since in such patients there is always chance and danger for epileptic seizures and cerebral ischaemia¹.

In our case, we also selected sevoflurane as the maintenance drug because it causes slight increases in cerebral blood flow and intracranial pressure at normocarbina, decreases cerebral oxygen requirements and produces no seizure activity, in comparison with other volatile anaesthetic agents²⁰. Its use in paediatric anaesthesia has been established, because it does not irritate the upper airway tract. providing continuous adequate anaesthesia depth. It has already

been suggested that in patients with moyamoya disease regional cortical blood flow levels may be decreased by inhalational anaesthesia³⁰. Despite this sole specific reference in literature, there are other studies already performed which suggest that the use of sevoflurane at 0.5, 1.0 and 1.5 minimum alveolar concentration (MAC) does not alter the cerebrovascular dynamics. Cho and colleagues reported that CO₂ reactivity and CBF were well maintained during 1.2 MAC sevoflurane anaesthesia, with and without nitrous oxide, in 14 volunteer patients at three different levels of PaCO₂³¹. Bedford et al demonstrated that transient hyperaemic response is well preserved during anaesthesia with sevoflurane up to 3.4% in oxygen³². Having in mind that generally sevoflurane does not compromise the cerebral vasculature balance and homeostasis, we chose sevoflurane for anaesthesia maintenance, without any adverse effects.

In conclusion, we report our experience from the anaesthetic management of a child, already operated for moyamoya disease, proceeding for inguinal hernia repair. Anaesthetic management focused on the maintenance of adequate CBF, normoventilation and normotension. This can be accomplished by minimally disturbing cerebrovascular resistance and maintaining cerebral perfusion pressure, to ensure adequate cerebral oxygenation. The use of LMA, under sevoflurane anaesthesia, contributed to the above goals, and proved to be safe and satisfactory, without causing any neurological adverse effects in the perioperative period.

REFERENCES

1. Gosalakkal JA: Moyamoya disease: a review. *Neurol India* 2002; 50: 6 – 10.
2. Hoffman HJ: Moyamoya disease and syndrome. *Clin Neurol Neurosurg* 1997; 99 Suppl 2: S 39 – S 44.
3. Matsushima Y, Inaba Y: Moyamoya disease in children and its surgical treatment. Introduction of a new surgical procedure and its follow – up angiograms. *Childs Brain* 1984; 11 (3): 155 – 170.
4. Gulden WD, Brandt M, Scherer R, Lawin P: Moyamoya syndrome in neuroanaesthesia. *Anasth Intensivther Notfallmed* 1987; 22: 33 – 36.
5. Susuki J, Takaku A: Cerebrovascular moyamoya disease: Disease showing abnormal net – like vessels in base of the brain. *Arch Neurol* 1969; 20: 288 – 299.
6. Scott MR: Surgery for moyamoya syndrome: Yes. *Arch Neurol* 2001; 58: 128 – 129.
7. Roach ES: Immediate surgery for moyamoya syndrome? Not necessarily. *Arch Neurol* 2001; 58: 30 – 31.

8. Sakamoto T, Kawaguchi M, Kurehara K et al: Postoperative neurologic deterioration following the revascularization surgery in children with moyamoya disease. *J Neurosurg Anesthesiol* 1998; 10: 37 – 41.
9. Golby AJ, Marks MP, Thompson RC: Direct and combined revascularization in pediatric moyamoya disease. *Neurosurgery* 1999; 45: 50 – 58.
10. Mugikura S, Takahashi S, Higano S, Shirane R, Sakurai Y, Yamada S: Predominant involvement of ipsilateral anterior and posterior circulations in moyamoya disease. *Stroke* 2002; 33: 1497 – 1500.
11. Robertson RL, Chavali RV, Robson CD et al: Neurologic complications of cerebral angiography in childhood moyamoya syndrome. *Pediatr Radiol* 1998; 28: 824 – 829.
12. Kansha M, Irita K, Takahashi S, Matsushima T: Anesthetic management of children with moyamoya disease. *Clin Neurol Neurosurg* 1997; 99 Suppl 2: S 110 – S113.
13. Futuya A, Matsukawa T, Ozaki M et al: Propofol anesthesia for cesarean section successfully managed in a patient with moyamoya disease. *J Clin Anesth* 1998; 10: 242 – 245.
14. Natori Y, Ikezaki K, Matsushima T et al: Angiographic moyamoya, its definition, classification and therapy. *Clin Neurol Neurosurg* 1997; 99 Suppl 2: S 168 – S 172.
15. Houkin K, Yoshimoto T, Kurodo S et al: Angiographic analysis of moyamoya disease – how does moyamoya disease progress? *Neurol Med Chir (Tokyo)* 1996; 36: 783 – 787.
16. Yonekawa Y, Ogata N, Kaku Y et al: Moyamoya disease in Europe, past and present status. *Clin Neurol Neurosurg* 1997; 99 Suppl 2: S 58 – S60.
17. Shetty – Alva N, Alva S: Familial moyamoya disease in Caucasians. *Pediatr Neurol* 2000; 23: 445 – 447.
18. Inoue TK, Ikezaki K, Sasazuki T et al: Linkage analysis of moyamoya disease on chromosome 6. *J Child Neurol* 2000; 15: 179 – 182.
19. Kuwabara Y, Ichiya Y, Sasaki M et al: Cerebral hemodynamics and metabolism in moyamoya disease – a positron emission tomography study. *Clin Neurol Neurosurg* 1997; 99 Suppl 2: S 74 – S 78.
20. Morgan EG Jr, Mikhail MS: Neurophysiology and anesthesia. In Morgan EG Jr, Mikhail MS (eds): *Clinical Anesthesiology*. Stamford: Appleton and Lange Publications, 1996; 477 – 490.
21. Brown SC, Lam AM: Moyamoya disease – a review of clinical experience and anaesthetic management. *Can J Anaesth* 1987; 34: 71 – 75.

22. Kurehara K, Ohnishi H, Touho H, Furuya H, Okuda T: Cortical blood flow response to hypercapnia during anaesthesia in moyamoya disease. *Can J Anaesth* 1993; 40: 709 – 713.
23. Yasuda C, Sumida T, Izima M, Nakarai E, Nemoto S, Aruga T: Anesthetic experience of a child with moyamoya disease. *Masui* 1989; 38: 809 – 812.
24. Tagawa T, Naritomi H, Mimaki T, et al: Regional cerebral blood flow, clinical manifestations, and age in children with moyamoya disease. *Stroke* 1987; 18: 906 – 910.
25. Sakamoto T, Kawaguchi M, Ohnishi H et al: Preoperative sedation for childhood moyamoya disease – clinical evaluation of rectally administered midazolam. *Masui* 1994; 43: 781 – 785.
26. Petty LA: Anesthetic management of a patient with moyamoya disease: a case report. *AANA J* 1993; 61: 277 – 281.
27. Ogawa A, Nakamura N, Yoshimoto T, et al: Cerebral blood flow in moyamoya disease. Part 2: Autoregulation and CO₂ response. *Acta Neurochir (Wien)* 1990; 105: 107 – 111.
28. Morgan EG Jr, Mikhail MS: Airway management. In Morgan EG Jr, Mikhail MS (eds): *Clinical Anesthesiology*. Stamford: Appleton and Lange Publications, 1996; 50 – 72.
29. Alexander J: Analgesic drugs. In: Aitkenhead AR, Rowbotham DJ, Smith G (eds): *Textbook of Anaesthesia*. Edinburgh: Churchill Livingstone Publishers, 2001; 211 –222.
30. Sato K, Shirane R, Kato M, Yoshimoto T: effects of inhalational anesthesia on cerebral circulation in moyamoya disease. *J Neurosurg Anesthesiol* 1999; 11: 25 – 30.
31. Cho S, Fujigaki T, Uchiyama Y et al: Effects of sevoflurane with and without nitrous oxide on human cerebral circulation. *Anesthesiology* 1996; 85: 755 – 760.
32. Bedforth N, Girling K, Harrison J et al: The effects of sevoflurane and nitrous oxide on middle cerebral artery blood flow velocity and transient hyperemic response. *Anesth Analg* 1999; 89: 170 – 174.