J Anesth (2014) 28:919–923
DOI 10.1007/s00540-014-1828-4

CLINICAL REPORT

Anesthesia for aortic reconstruction in a child with PHACE syndrome

Tatsuyuki Imada · Ryu Okutani · Yutaka Oda

Received: 30 December 2013 / Accepted: 25 March 2014 / Published online: 18 April 2014

Abstract PHACE syndrome is a neurocutaneous syndrome characterized by the association of large cutaneous hemangiomas and the cardiac and cerebral vascular anomalies. We report a 6-year-old female with PHACE syndrome presented with left facial hemangiomas, cystic lesion in the cerebral posterior fossa, coarctation of the aorta, aplasia of the left vertebral artery and stenosis of the left internal carotid artery. Surgical repair of the aorta with left heart bypass under beating heart was scheduled. We monitored regional cerebral oxygen saturation (rSO_2) with infrared spectroscopy in order to detect cerebral hypoperfusion. A decrease of rSO_2 ipsilateral to the cerebrovascular anomalies occurred during anastomosis of the aorta, which was treated by reducing the flow rate of left heart bypass and by increasing the inhalational oxygen concentration. As children with PHACE syndrome are frequently accompanied with cerebrovascular anomalies and at a risk of cerebral hypoperfusion, prevention of cerebral hypoperfusion is crucially important during general anesthesia.

Keywords Coarctation of the aorta · Dandy-Walker syndrome · Left heart bypass · PHACE syndrome · Regional cerebral oxygen saturation

Introduction

PHACE syndrome[1] is a coined acronym consisting of the initial letters of the following features: posterior fossa brain malformations, hemangiomas on face, anomaly of the cerebral arteries, coarctation of the aorta, and eye anomaly [1, 2]. Children with this syndrome are often required to undergo surgery for aortic arch anomalies in early childhood [2, 3]. As they are frequently accompanied with multiple stenotic and occlusive anomalies in the cerebral arteries and at a risk of arterial ischemic stroke [2, 4], prevention of cerebral hypoperfusion is crucially important during anesthesia. Despite these challenging conditions, there are only few reports describing anesthesia for patients with PHACE syndrome [5]. Here we report a child with PHACE syndrome accompanied with multiple aortic and cerebrovascular anomalies undergoing aortic reconstruction under left heart bypass.

Case report

A 6-year-old female (height 108 cm; weight 21 kg) presented with complaints of headache and dyspnea on effort. She was delivered via Caesarean section at 34 weeks 5 days with an ultrasonic prenatal diagnosis of Dandy-Walker syndrome, and was transferred to our hospital for further examination. A diagnosis of PHACE syndrome was made based on facial hemangiomas in all left trigeminal areas, cerebral posterior fossa lesion and aortic tortuosity. Either angiography or three-dimensional computed tomography (3D-CT) did not reveal aortic coarctation. At 28 days of age, cyst-peritoneal shunting was performed for hydrocephalus under general anesthesia uneventfully. She had normal physical, and motor development without symptoms of cerebral ischemia, but with slight mental

T. Imada · R. Okutani · Y. Oda (✉)
Department of Anesthesiology, Osaka City General Hospital and Children's Hospital, 2-13-22, Miyakojima-hondori, Miyakojimaku, Osaka 534-0021, Japan
e-mail: odayou@msic.med.osaka-cu.ac.jp

[1] Online Mendelian Inheritance in Man database No. 606519.

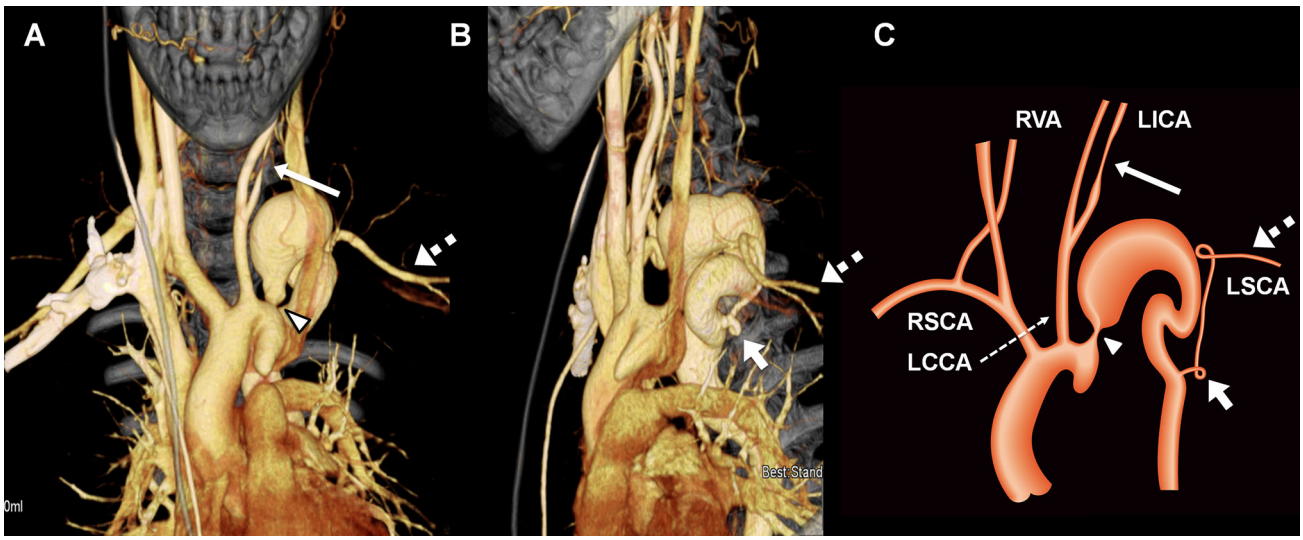

Fig. 1 Three-dimensional computed tomography imaging of the aorta from **a** anterior, **b** left lateral view and **c** schematic diagram. Coarctation distal to the branching portion of the left common carotid artery (*arrow head*, **a** and **c**) with aneurysmal dilation and extended tortuosity of the aortic arch. The left subclavian artery arises from the aorta distal to aneurysmal dilation with a small loop at the proximal portion (*short and wide arrow*, **b** and **c**), ascends to the level of the aortic arch and extends to the left upper arm (*wide and dashed arrow*, **a**–**c**). Stenosis of the left internal carotid artery (*long and narrow arrow*, **a** and **c**). *RSCA* right subclavian artery, *RVA* right vertebral artery, *LCCA* left common carotid artery, *LICA* left internal carotid artery, *LSCA* left subclavian artery

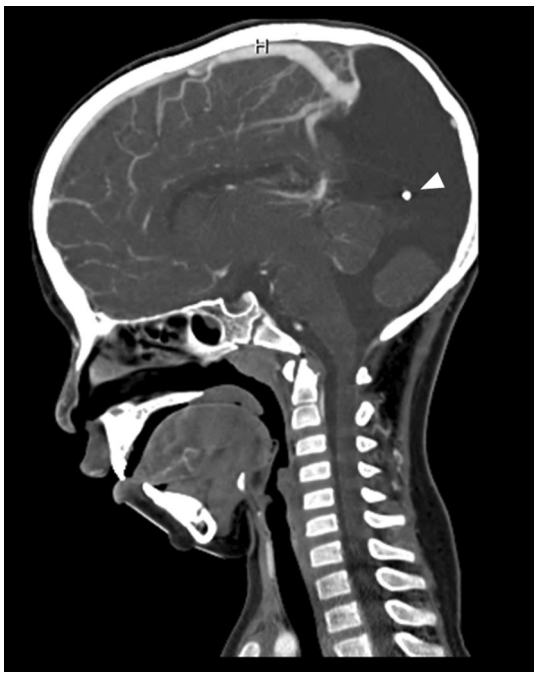

Fig. 2 Magnetic resonance imaging of the brain. Cystic lesion in the posterior fossa with a cyst-peritoneal shunt tube (*arrow head*)

retardation, and has been followed up by neurologists with no medication.

On admission, physical examination showed left facial hemangiomas with regression compared with those during the neonatal period, and a faint pulse in the legs with no neurological deficits. Blood pressure was 126/96, 106/79 and 95/57 mmHg on the right and left arms, on the right leg, respectively. Transthoracic echocardiography demonstrated satisfactory left ventricular contractility with an ejection fraction of 63 %, absence of intracardiac malformation, and presence of stenosis in the distal aortic arch. The maximum and mean pressure gradient across the stenotic lesion was 88 and 55 mmHg, respectively. 3D-CT showed marked tortuosity, coarctation of the aortic arch distal to the branching portion of the left common carotid artery, and aneurysmal dilation distal to the coarctation without aberrant origin of the subclavian arteries (Fig. 1). It also revealed the absence of the left vertebral artery and stenosis of the left internal carotid artery with collateral through the anterior communicating artery. Magnetic resonance imaging showed a massive cystic lesion in the posterior fossa (Fig. 2). Surgical reconstruction of the aortic arch was planned.

After premedication with midazolam 9 mg orally, standard monitoring was started in the operating room. Blood pressure on the right arm was 95/43 mmHg. Electrodes for monitoring regional cerebral oxygen saturation (rSO_2) using near-infrared spectroscopy (INVOS^{TM} 5100, Covidien, Mansfield, MA, USA) were applied immediately after induction of general anesthesia with sevoflurane, and the baseline values on the right and left sides were 71 and 70 %, respectively. Limited space in her forehead prevented us from applying additional electrodes for monitoring bispectral index. After administration of rocuronium

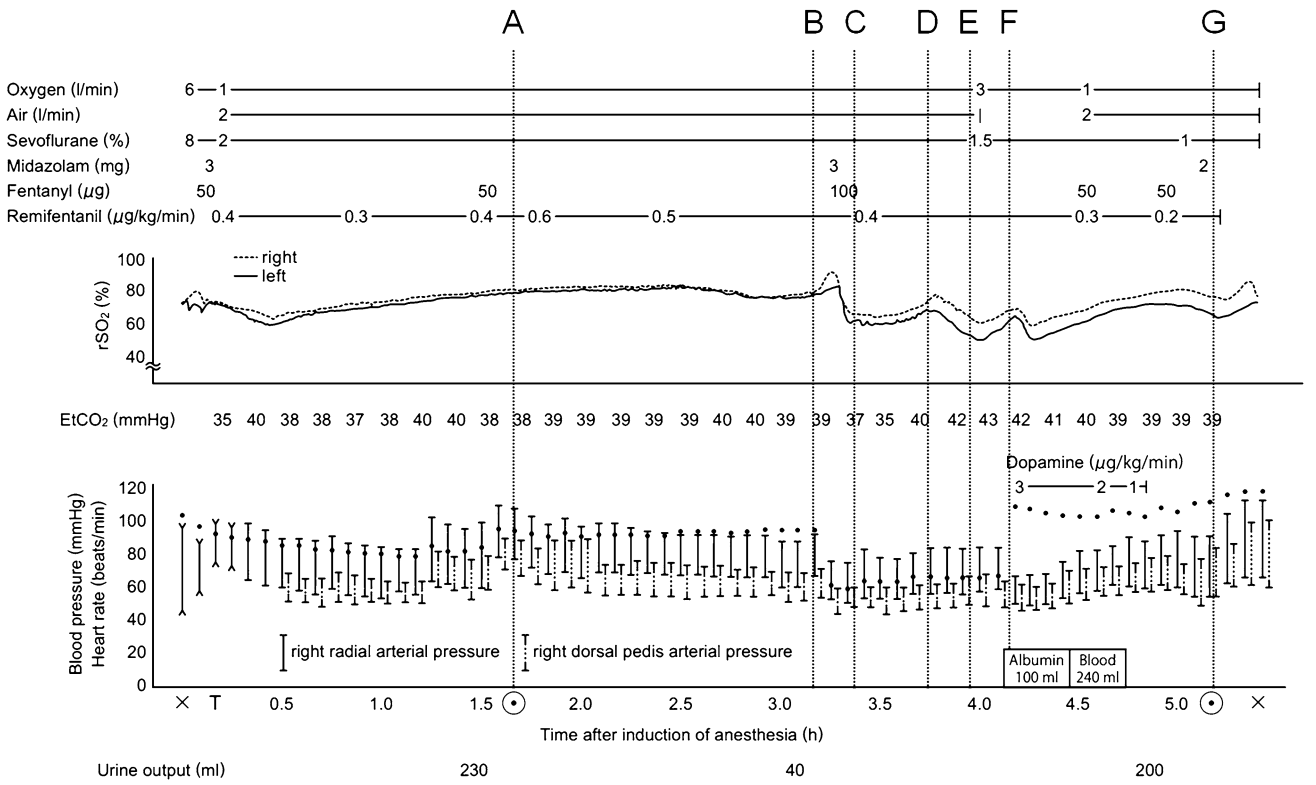

Fig. 3 Anesthesia record showing right radial and dorsal pedis arterial pressure, end-tidal carbon dioxide tension and regional cerebral oxygen saturation (rSO_2). *Solid curve* left-sided rSO_2, *dashed curve* right-sided rSO_2. *Vertical lines* indicate start of **a** surgery, **b** left heart bypass at a flow rate of 500 ml/min, **c** proximal aortic anastomosis, **d** distal aortic anastomosis, **e** reduction of the flow rate of left heart bypass to 300 ml/min, **f** completion of reconstruction of the aortic arch and declamp of the descending aorta, **g** end of surgery. Peripheral oxygen saturation was maintained at 100 % during anesthesia

15 mg, the trachea was intubated, she was ventilated with 45 % oxygen. No hemangiomas were detected during laryngoscopy and tracheal intubation. Intraarterial catheters were inserted into the right radial and dorsal pedis arteries, and a central venous catheter was placed in the right internal jugular vein. Anesthesia was maintained with sevoflurane, fentanyl, and remifentanil.

After a left fourth intercostal thoracotomy and small-dose systemic heparinization (100 u/kg), a 22 F venous cannula and a 12 F arterial cannula were inserted into the left atrial appendage and the descending aorta, respectively. Left heart bypass was performed with a centrifugal pump at a flow rate of 500 ml/min for maintaining mean radial arterial pressure above 60 mmHg and mean dorsal pedis arterial pressure exceeding 50 mmHg. We tried to maintain rSO_2 above 50 % based on previous reports [5, 6]. Blood temperature was maintained at 35.5 °C. After ligating both proximal and distal sides of the aneurysmal dilation of the aorta, an expanded polytetrafluoroethylene graft (size 14 mm) was anastomosed end to side of the aortic arch distal to the branching portion of the left common carotid artery, then anastomosed end to side of the aortic arch proximal to the branching of the left subclavian artery.

Bilateral rSO_2 decreased by approximately 20 % after starting left heart bypass, in accordance with a decrease of mean radial arterial pressure from 76 mmHg to 60 mmHg (Fig. 3; Table 1). rSO_2 on the left side further decreased below 50 % during anastomosis of the distal side of the graft without further decrease of radial arterial pressure. We attempted to increase cerebral blood flow by reducing the flow rate of left heart bypass to 300 ml/min. We also increased the inhalational oxygen concentration from 45 to 100 %, while maintaining end-tidal carbon dioxide tension above 40 mmHg. Bilateral rSO_2 gradually increased and the differences between right and left sides decreased. The mean dorsal pedis arterial pressure remained above 50 mmHg during left heart bypass. After completion of aortic reconstruction and declamp of the descending aorta, we started continuous infusion of dopamine at 3 μg/kg/min in order to increase blood pressure. Surgery was completed uneventfully and the patient entered the intensive care unit under tracheal intubation. Duration of surgery, anesthesia and left heart bypass was 3 h 30 min, 5 h 24 min and 51 min, respectively. She was extubated on the postoperative day 1, and discharged home on the day 11 without any neurological complications. She is 7 year

Table 1 Hemodynamic parameters and regional cerebral oxygen saturation during anesthesia

Time points	A	B	C	D	E	F	G
Right radial arterial pressure (mmHg)	109/78/86	90/67/76	76/53/60	84/60/66	85/58/66	70/55/60	93/56/67
Right dorsal pedis arterial pressure (mmHg)	90/65/76	71/54/60	60/46/51	62/49/53	64/46/51	62/46/51	82/57/63
Heart rate (beats/min)	96	95	64	66	65	110	113
Central venous pressure (mmHg)	6	6	4	3	6	7	9
Peripheral oxygen saturation (%)	100	100	100	100	100	100	100
Right rSO_2 (%)	80	80	66	73	60	70	80
Left rSO_2 (%)	78	78	62	67	50	64	72
End-tidal carbon dioxide tension (mmHg)	38	39	37	40	43	42	39
Pharyngeal temperature (°C)	36.0	35.8	35.5	35.5	35.5	36.2	36.2
Hemoglobin content (g/dl)	10.7			7.5			8.4

Arterial blood pressure is expressed as systolic/diastolic/mean values. Time points *A* start of surgery, *B* start of left heart bypass at a flow rate of 500 ml/min, *C* start of proximal aortic anastomosis, *D* start of distal aortic anastomosis, *E* reduction of the flow rate of left heart bypass to 300 ml/min, *F* completion of reconstruction of the aortic arch and declamp of the descending aorta, *G* end of surgery, rSO_2 regional cerebral oxygen saturation

of age, with no sensory or motor deficits except slight mental retardation, free from complaints of headache and dyspnea.

Discussion

The concept of PHACE syndrome was developed by Frieden in 1996 [1]. Hemangiomas, found in almost all affected patients, tend to be large (>5 cm in diameter) and segmental [2]. They usually grow during infancy and regress thereafter [2]. Supra and subglottic hemangiomas are also reported [7]. Dandy-Walker malformation is the most common intracranial lesion [2]. Coarctation of the aorta is the most common cardiovascular lesion, often associated with perivascular tortuosity and aneurysmal dilation [2, 3]. Cardiac and cerebrovascular anomalies and cutaneous hemangiomas are most often ipsilateral [3]. Despite the diagnosis of PHACE syndrome made on the next day after birth in the present case, coarctation of the aorta was not detected until the age of 6 years, possibly resulting from developed collateral circulation to the lower part of the body.

Children with PHACE syndrome are at an increased risk of cerebral ischemia secondary to hypoperfusion [4] and of possible airway obstruction due to airway hemangioma [7]; however, there are only few reports describing anesthesia in those patients [5]. As preoperative assessment revealed aplasia of the left vertebral artery and stenosis of the left internal carotid artery, protection of the brain as well as spinal cord from ischemia was required during anesthesia. We used left heart bypass to prevent spinal ischemia, which is effective for preventing spinal ischemia during surgery of aortic coarctation [8]. However, shift of blood from the systemic circulation to the bypass flow may lead cerebral hypoperfusion. We maintained mean right radial arterial pressure above 60 mmHg for maintaining cerebral blood flow, based on the level before induction of general anesthesia. We also monitored rSO_2. Although interventions based on rSO_2 do not necessarily protect patients from brain injury [9] or monitoring of rSO_2 is not covered by public health insurance in our country, it is useful for detecting cerebral hypoperfusion during cerebral vascular surgery and repair of aortic coarctation [10, 11]. As the space for applying the electrodes was limited and rSO_2 is more sensitive for detecting cerebral hypoperfusion than bispectral index [5], we selectively monitored rSO_2.

An abrupt decrease in bilateral rSO_2, probably due to hemodilution as reported previously [5, 12], was observed after initiating left heart bypass (Fig. 3; Table 1). Interventions to counter cerebral ischemia as shown by further decrease of rSO_2 on the left side were undertaken that involved a reduction of the flow rate of left heart bypass in order to increase the cerebral blood flow and oxygen supply, while maintaining mean dorsal pedis blood pressure above 50 mmHg, and an increase of inhalational oxygen concentration, resulting in an increase of rSO_2.

Javault et al. [5] reported anesthesia for a child with PHACE syndrome undergoing thoracic aorto-aortic bypass graft under cardiopulmonary bypass. They also detected a remarkable decrease of rSO_2 immediately after declamping the descending aorta, probably resulting from decreased blood pressure and administered a vasopressor for increasing blood pressure. However, rSO_2 remained decreased until the end of cardiopulmonary bypass. In sharp contrast, a decrease of rSO_2 was successfully treated by reducing the flow rate of left heart bypass and by increasing inhalational oxygen concentration. Our

intervention would have been more beneficial than using vasopressors, which may lead to hypoperfusion of the organs.

In conclusion, we performed anesthesia for surgical repair of the aorta in a child with PHACE syndrome. Given the possibility of cerebral ischemia due to cerebrovascular anomalies during anesthesia, prevention of cerebral hypoperfusion is important.

Conflict of interest Financial support was solely from departmental sources, and there are no potential conflicts of interest.

References

1. Frieden IJ, Reese V, Cohen D. PHACE syndrome. The association of posterior fossa brain malformations, hemangiomas, arterial anomalies, coarctation of the aorta and cardiac defects, and eye abnormalities. Arch Dermatol. 1996;132:307–11.
2. Metry D, Heyer G, Hess C, Garzon M, Haggstrom A, Frommelt P, Adams D, Siegel D, Hall K, Powell J, Frieden I, Drolet B, Conference PSR. Consensus statement on diagnostic criteria for PHACE syndrome. Pediatrics. 2009;124:1447–56.
3. Rao RP, Drolet BA, Holland KE, Frommelt PC. PHACES association: a vasculocutaneous syndrome. Pediatr Cardiol. 2008;29:793–9.
4. Drolet BA, Dohil M, Golomb MR, Wells R, Murowski L, Tamburro J, Sty J, Friedlander SF. Early stroke and cerebral vasculopathy in children with facial hemangiomas and PHACE association. Pediatrics. 2006;117:959–64.
5. Javault A, Metton O, Raisky O, Bompard D, Hachemi M, Gamondes D, Ninet J, Neidecker J, Lehot JJ, Cannesson M. Anesthesia management in a child with PHACE syndrome and agenesis of bilateral internal carotid arteries. Paediatr Anaesth. 2007;17:989–93.
6. Cho H, Nemoto EM, Yonas H, Balzer J, Sclabassi RJ. Cerebral monitoring by means of oximetry and somatosensory evoked potentials during carotid endarterectomy. J Neurosurg. 1998;89:533–8.
7. Smith DS, Lee KK, Milczuk HA. Otolaryngologic manifestations of PHACE syndrome. Int J Pediatr Otorhinolaryngol. 2004;68:1445–50.
8. Fiore AC, Ruzmetov M, Johnson RG, Rodefeld MD, Rieger K, Turrentine MW, Brown JW. Selective use of left heart bypass for aortic coarctation. Ann Thorac Surg. 2010;89:851–6.
9. Ghosh A, Elwell C, Smith M. Review article: cerebral near-infrared spectroscopy in adults: a work in progress. Anesth Analg. 2012;115:1373–83.
10. Joshi RK, Motta P, Horibe M, Mossad E. Monitoring cerebral oxygenation in a pediatric patient undergoing surgery for vascular ring. Paediatr Anaesth. 2006;16:178–81.
11. Farouk A, Karimi M, Henderson M, Ostrowsky J, Siwik E, Hennein H. Cerebral regional oxygenation during aortic coarctation repair in pediatric population. Eur J Cardiothorac Surg. 2008;34:26–31.
12. Hayashida M, Kin N, Tomioka T, Orii R, Sekiyama H, Usui H, Chinzei M, Hanaoka K. Cerebral ischaemia during cardiac surgery in children detected by combined monitoring of BIS and near-infrared spectroscopy. Br J Anaesth. 2004;92:662–9.