Abstract
Pheochromocytoma is rarely seen under the age of 8 years and the anaesthesia for pheochromocytoma surgery is challenging. We present the perioperative management of a 5-year-old boy with intestinal obstruction and two episodes of cerebral haemorrhage who presented for pheochromocytoma resection and enterolysis. Important issues in the management of this patient included choice of vasoactive agents and anaesthetics, control of blood pressure and intracranial pressure, preoperative fluid management, and monitoring technique. Imbalance of fluid and electrolyte disturbance as well as a potentially inevitable drop in blood pressures after complete excision of tumour further complicate patient care. Knowledge of different medical issues involved in this case and their potential impact on anaesthetic management is paramount for safe perioperative patient care.

Keywords: General anaesthesia, Cerebral haemorrhage, Intraoperative monitoring, Paediatrics, Pheochromocytoma.

Introduction
Characterized by high secretion of catecholamines, pheochromocytoma is a tumour that originates from pheochromocytes in the medullar portion of the adrenal glands. This tumour is very rare in children. Only 5% of all pheochromocytomas have been reported in children and the incidence is very low below the age of 8 years.1 While surgical excision is the definitive treatment, the anaesthesia management presents a challenge, since usually neither preoperative preparations nor management during general anaesthesia can totally prevent haemodynamic fluctuations during surgical manipulation or after removal of the tumour.2 In children, cases regarding cerebral haemorrhage caused by pheochromocytoma have been rarely reported so far. No report on pheochromocytoma combined with intestinal obstruction in a child was found in literature. Given the rarity of pheochromocytoma with cerebral haemorrhage and intestinal obstruction in a child, there is no published case on its anaesthetic management. Here, we describe the perioperative management of pheochromocytoma resection and enterolysis in a 5-year-old boy with cerebral haemorrhage and intestinal obstruction. The patient’s guardian, his father, gave written consent for publication of this report. An ethical approval of this case report was obtained from our hospital.

Case Report
A 5-year-old boy presenting with a history of sweating, headaches and polyuria for 4 months was admitted to local hospital in March, 2015. Medical examination revealed tachycardia (173 beats/min), hypertension (215/142 mmHg), haemorrhage of the left frontal lobe, and hypertrophy of the left ventricle (ejection fraction: 61%). Blood pressure (BP) and intracranial pressure (ICP) were controlled by sodium nitroprusside and mannitol. Removal of intracranial haematoma and decompressive craniectomy were performed under general anaesthesia. He was transferred to our hospital’s paediatric ward on 16 April, 2015 since headaches and vomiting worsened. Brain CT showed cerebral haemorrhage of the right temporal lobe (Figure 1-A). Thoracic and abdominal CT showed a 37×38 mm mass in the left adrenal gland (Figure 1-B). Urinary 24 hour vanillylmandelic acid (VMA) was 192.62 µmol (reference range <68.6 µmol). The levels of blood glucose, serum potassium and serum calcium were 131.4 mg/dL, 11.3 mg/dL and 11.24 mg/dL respectively.

After admission, heart rate (HR), BP, and ICP were controlled by esmolol, urapidil, phenoxybenzamine and mannitol. The patient complained of abdominal distension and pain, which was diagnosed as intestinal obstruction, indicated by abdominal plain X-ray films (Figure 1-C).

When BP and HR were controlled within 120-150/80-100 mmHg and 110-140 beats/min with antihypertensive drugs, the surgical procedures were performed. Anaesthesia was induced with midazolam 0.1 mg/kg, sufentanil 1 µg/kg, propofol 2 mg/kg and cisatracurium 0.2 mg/kg. Intubation was done with a bispectral index...
BIS value of 57, followed by a temporary increase in HR and BP (Figure-2). Anaesthesia was maintained with remifentanil 0.1-0.3 μg/(kg.min) and propofol 6-9 mg/(kg.h) intravenously. Central venous pressure (CVP), invasive arterial pressure and stroke volume variation (SVV) were monitored.

The first surgical procedure was resection of the tumour. Haemodynamic fluctuations occurred during the manipulation (Figure-2), which was controlled with repeated intravenous injections of esmolol 0.1 mg/kg. Antihypertensive drugs were discontinued once the tumour was removed (Figure 1-D). There was a precipitous fall of BP and CVP in the initial period of enterolysis, together with an increase of SVV (Figure 2). Managed with infusions of phenylephrine, norepinephrine and epinephrine, the haemodynamics was relatively stable with about 10-20% deviation from the baseline. Guided by monitor results, intraoperative fluid was infused accurately. Blood gases and oxygenation were normal during the surgery except a progressive decrease of serum potassium, which was treated with potassium chloratum injection. The anaesthesia lasted 3 hours and 45 minutes. Estimated blood loss was 200 mL and urine output was 270 mL. Transfusion included crystalloid 630 mL, colloid 200 mL, concentrated blood cells 100 mL and plasma 200 mL. Post-operative analgesia was achieved by oxycodone 2 mg intravenously.

Except SVV and BIS, monitors were continued in the intensive care unit (ICU) where norepinephrine 0.05 μg/(kg.min) was required for maintenance of BP temporarily and potassium chloratum injection was required to treat hypokalaemia. The child was returned to paediatric ward with stable haemodynamics and normal serum potassium level on 4th day postoperatively. The pathological result demonstrated pheochromocytoma and urinary 24 hr VMA decreased to 31.7 μmol (reference range < 68.6 μmol) on 10th day postoperatively. The child was discharged home on 14th postoperative day.
Discussion

Children with pheochromocytoma presenting as intracranial haemorrhage was only reported in a case of two Taiwanese children. In this case, the patient suffered 2 episodes of cerebral haemorrhage. Mannitol was continued and fluid infusion was deficient in case of cerebral hernia. Additionally, the complexity of fluid management was augmented by presenting of intestinal obstruction. These pathological conditions confer upon more risk of inevitable drop in blood pressures following excision of the tumour. Close monitoring and precise control of haemodynamics is necessary. CVP, reflecting right atrial pressure, was found not a reliable predictor of fluid responsiveness in the paediatric population from recent studies. As a predictor of fluid responsiveness in mechanically ventilated adults in sinus rhythm, SVV has been demonstrated to be a reliable parameter in children. We monitored both SVV and CVP during the procedure, and they had a good correlation, which is consistent with the recent study. BIS was monitored to achieve stable haemodynamics and minimize the use of additional vasoactive agents.

In such a case, drug selection should be made carefully to achieve pharmacological control of haemodynamics. Urapidil was selected taking its advantage of no obvious influence on ICP. Tachycardia as a consequence of elevated catecholamine was treated with esmolol. The intraoperative haemodynamic elevations were well treated by the combined use of urapidil and esmolol. Sodium nitroprusside, inducing cerebrovascular dilatation and ICP increase, was just prepared for unmanageable hypertension. The hypotension in the initial stage of enterolysis can be a consequence of blood volume depletion, and long-acting non-specific α-adrenergic blocking agents.

Hyperglycaemia and hypokalaemia occurred in the child. Besides abnormal catecholamines secretion, long-term fasting and continuous gastrointestinal decompression also induce hypokalaemia. Pheochromocytoma, irregular diet, complete bed rest and hypokalaemia might contribute to the intestinal obstruction in this patient. So, it should be emphasized to monitor perioperative metabolic disorder in patients undergoing pheochromocytoma resection.

Conclusion

We describe the anaesthetic management for pheochromocytoma resection and enterolysis in a 5-year-old boy with cerebral haemorrhage and intestinal obstruction. Proper and adequate preoperative preparation through using of vasoactive drugs and restoration of blood volume, appropriate anaesthesia choice and close perioperative monitoring guarantee good haemodynamic control, resulting in a favourable outcome.

Conflict of Interest: None.

Disclaimer: The manuscript has not been presented in a conference and is not under consideration for publication in any other journal.

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References