Anaesthetic management of phaeochromocytoma associated with tricuspid atresia

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The anaesthetic management of a patient with phaeochromocytoma, tricuspid atresia and pulmonary vascular stenosis is reported. The patient received no preoperative preparation with adrenergic blockers. Anaesthesia was induced and maintained with fentanyl, diazepam and sevoflurane. Intraoperative blood pressure was controlled with sodium nitroprusside, sevoflurane, phentolamine, and propranolol. For hypotension after resection of the tumour norepinephrine was required. This patient did not have a systemic to pulmonary shunt procedure performed, so the maintenance of pulmonary blood flow in the presence of haemodynamic instability during operation for phaeochromocytoma was a major concern. Monitoring of oxyhaemoglobin saturation (SpO₂) with a pulse oximeter was considered to be useful because SpO₂ may reflect pulmonary flow. During serious haemodynamic disturbances due to the manipulation of the tumour, the heart rate was inversely correlated with SpO₂, but the relationship between mean arterial pressure and SpO₂ was weak. Therefore, control of heart rate appeared to be more important than control of blood pressure in this case.

La conduite anesthésique d'un patient atteint de phaeochromocytome, atresie tricuspide et de sténose pulmonaire est reportée. Le patient n'a pas reçu de traitement pré-opératoire avec des bloqueurs adrénergiques. L'anesthésie fut induite et maintenue avec du fentanyl, diazepam et sevoflurane. La pression artérielle per-opératoire fut contrôlée avec du nitro-

pussiate, servoflurane, phentolamine et propranolol. Pour l'hypotension après la résection de la tumeur la norepinephrine fut requise. Ce patient n'a pas eu de Shunt systémique pulmonaire et ainsi le maintien du flot sanguin pulmonaire en présence d'une instabilité hémodynamique durant l'opération représentait un souci majeur. La surveillance de la saturation de l'oxyhaémoglobine (SpO₂) avec un saturomètre de pouls fut considérée utile car la SpO₂ peut refléter le flot sanguin pulmonaire. Lors des altérations hémodynamiques sévères dues à la manipulation de la tumeur, la fréquence cardiaque fut inversement corrélée à la SpO₂ et la relation entre la pression artérielle moyenne et la SpO₂ était faible. Ainsi, le contrôle de la fréquence cardiaque nous a paru plus important que le contrôle de la pression artérielle dans ce cas.

The management of phaeochromocytoma is a challenge for anaesthetists.^{1,2} According to recent reports,³⁻⁸ it has been performed successfully with a variety of inhaled and intravenous anaesthetics, adrenergic blockers, vasodilators, calcium channel blockers or combined administration of these agents. However, the coexistence of a serious cardiovascular abnormality with phaeochromocytoma may result in unusual difficulty in intraoperative management.^{9,10}

This is a report of the anaesthetic management of a patient with phaeochromocytoma associated with tricuspid atresia and pulmonary vascular stenosis. A palliative shunt to the pulmonary artery was not created. Therefore, the maintenance of pulmonary blood flow in the presence of serious haemodynamic change during operation for resection of a phaeochromocytoma was considered to be essential.¹⁰ We used continuous monitoring of oxyhaemoglobin saturation (SpO₂) with a pulse oximeter to estimate pulmonary blood flow and examined which haemodynamic variables affected the balance between systemic and pulmonary blood flow.

Case report

A 16-yr-old boy, weighing 55 kg, was admitted for evaluation before Fontan operation to correct tricuspid atresia and pulmonary vascular stenosis. Cyanosis oc-

Key words

SURGERY: phaeochromocytoma;

COMPLICATIONS: tricuspid atresia

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curred on the first day of his life. At the age of three years, cardiac catheterization showed tricuspid atresia, ventricular septal defect, atrial septal defect and pulmonary vascular stenosis. As there was no sign of heart failure or cyanotic spells, he was given no medication and was seen regularly as an outpatient. On admission, physical examination revealed no sign of cardiac failure, but unaccountable hypertension (systolic pressure 160–180 mmHg, diastolic pressure 100–110 mmHg) was observed. Cardiac catheterization at this time demonstrated severe pulmonary vascular stenosis as well as tricuspid atresia, a large ventricular septal defect and an atrial septal defect. Left ventricular and aortic pressures were 136/0 mmHg and 138/104 mmHg, respectively, but pulmonary arterial pressure was not elevated (10/7 mmHg). The ratio of pulmonary and systemic arterial pressure (P_p/P_s), the ratio of pulmonary and systemic blood flow (Q_p/Q_s) and the ratio of pulmonary and systemic resistance (R_p/R_s) were 0.1, 0.9 and 0.1, respectively. Ventricular wall motion was well preserved. The ECG revealed sinus rhythm, left ventricular hypertrophy and incomplete right bundle branch block. The haematocrit and haemoglobin concentration were markedly elevated (68.5% and 20.6 mg · dl⁻¹, respectively) and the arterial pH, PaO₂, PaCO₂ and oxygen saturation were 7.45, 42.6 mmHg, 40.0 mmHg and 80% while breathing room air, respectively. Further laboratory examination for hypertension indicated the presence of phaeochromocytoma; plasma norepinephrine and epinephrine concentrations were 8300 pg · ml⁻¹ (normal values; 196–490 pg · ml⁻¹) and 33 pg · ml⁻¹ (13–61 pg · ml⁻¹) respectively, and urine norepinephrine excretion was 944 µg · day⁻¹ (30–120 µg · day⁻¹). The diagnosis of phaeochromocytoma was confirmed by abdominal computerized axial tomography which showed a 35 × 28 mm mass above to the left renal pole. Surgery for the phaeochromocytoma was scheduled before the Fontan operation.

The patient received no preoperative preparation with adrenergic blocking agents. He was given premedication with scopolamine 0.4 mg, *im* and diazepam 10 mg, *po*, 55 min and two hours before anaesthesia, respectively. Before induction of anaesthesia, monitoring included an ECG (lead II), pulse oximeter (Nellcor N 1000®, Hayward, CA), and an automated blood pressure monitor (Dinamap®, Critikon, Tampa, FL). On arrival in the operating room, his blood pressure was 170/110 mmHg. An 18-G catheter was inserted in a left peripheral vein and the right radial artery was cannulated under local anaesthesia for both pressure monitoring and blood sampling. Anaesthesia was induced with fentanyl, 300 µg, diazepam 5 mg, and vecuronium 10 mg was given to produce muscle relaxation. As the blood pressure was still elevated, inhalation of sevoflurane (2%) was added.

FIGURE 1 Intraoperative recordings of blood pressure, heart rate and oxyhaemoglobin saturation (SpO₂). A = initiation of the operation; B = resection of the tumour.

When the arterial blood pressure decreased to 105/76 mmHg, the trachea was intubated and this was achieved without marked elevation of the blood pressure. Anaesthesia was maintained with fentanyl and sevoflurane with air and oxygen (FiO₂ 0.5–1.0) and muscular relaxation was achieved with supplementary doses of vecuronium. A triple lumen catheter was inserted via the right internal jugular vein for the monitoring of central venous pressure (CVP) and for the administration of drugs. Sodium nitroprusside (SNP) was administered *iv* by continuous infusion of 0.3–0.5 µg · kg⁻¹ · min⁻¹. Before manipulation of the tumour, a total of 1200 ml blood was lost and 3850 ml of crystalloid and colloid solution were administered over 120 min, because hyperviscosity of blood might exaggerate heart failure. As a result, haematocrit was reduced from 63% to 54%, but blood pressure and CVP were not changed. The blood lost was collected for autotransfusion. Both the right femoral vein and artery were exposed before exploration of the abdomen in order to prepare the ECLA (extracorporeal lung assist) in the event of an anoxic spell due to a decrease in pulmonary blood flow. The blood pressure and heart rate changes that occurred during tumour manipulation are shown in Figure 1. Intraoperative blood pressure was controlled with SNP and sevoflurane, but the blood pressure elevation and increase in heart rate, due to surgical manipulation of the tumour, were managed with bolus injections of phentolamine (0.5–1.0 mg) or propranolol (0.1 mg). During surgical manipulation, SpO₂ and heart rate were inversely

FIGURE 2 Correlation between mean arterial pressure and SpO_2 during tumour manipulation.

related, but the relationship between SpO_2 and mean arterial pressure was weak (Figure 2). However, there was a strong relationship between heart rate and SpO_2 (Figure 3). The SpO_2 was maintained between 72 and 86% throughout the operation. After ligation of the adrenal vein, a decrease in blood pressure occurred, which was managed with cessation of SNP infusion, administration of a plasma expander, and continuous infusion of dopamine with $3.0\text{--}5.0 \mu\text{g}\cdot\text{kg}^{-1}\cdot\text{min}^{-1}$. However, an increase in heart rate following dopamine administration resulted in a reduction of SpO_2 , so dopamine was replaced by norepinephrine ($0.06\text{--}0.3 \mu\text{g}\cdot\text{kg}^{-1}\cdot\text{min}^{-1}$). The blood loss of 660 ml was managed by 400 ml of autotransfusion. During the procedure, haemodynamic variables changed little. No arrhythmia was observed and severe hypoxia requiring the use of ECLA did not occur. Intermittent measurements of arterial pH, PaO_2 and PaCO_2 were in the range of 7.33–7.37, 42.9–49.6 mmHg, and 45.0–48.8 mmHg, respectively. The patient was taken to the intensive care unit where the cardiac support by norepinephrine was maintained for three hours. He was recovered from anaesthesia uneventfully, and the tracheal tube was removed on the day following surgery. He was discharged from the intensive care unit on the second postoperative day. In order to measure plasma epinephrine and norepinephrine concentrations, a 6 ml arterial blood sample was collected before induction of anaesthesia, after tracheal intubation, during exploration of the abdomen, during manipulation of the tumour and after its resection.

FIGURE 3 SpO_2 was inversely correlated with heart rate during tumour manipulation.

Furthermore, a sample of blood from autotransfusion was analyzed for catecholamines. Plasma concentrations of catecholamines were determined by the method described by Yamatodani and Wada,¹¹ which was based on the trihydroxyindole reaction. The results of the analysis are shown in the Table. Both norepinephrine and epinephrine concentrations increased markedly during manipulation of the tumour compared with before induction. However, the concentrations during autotransfusion were similar to those during exploration of the abdomen.

Discussion

This case report describes the management of a patient with tricuspid atresia and pulmonary vascular stenosis who developed a pheochromocytoma. Pheochromocytoma is a rare tumour in the paediatric population¹² and the incidence of tricuspid atresia is about 1% in children with congenital heart disease.¹³ Although a correlation between cyanotic congenital heart disease and pheochromocytoma has been reported,¹⁴ the chance of an anaesthetist being faced with a similar case is rare. In the present case, pulmonary blood flow depended on blood flow through VSD from left ventricle. Decreased systemic vascular resistance should decrease blood flow through VSD, resulting in a decrease of pulmonary blood flow and hypercontractility of ventricle secondary to tachycardia or hypertension might also decrease pulmonary blood flow because of pulmonary stenosis. Therefore, haemody-

TABLE Plasma norepinephrine and epinephrine concentrations before and during resection of phaeochromocytoma (pg ml^{-1})

	Norepinephrine	Epinephrine
Before induction	5772	72
After intubation	5592	216
Exploration of the abdomen	2919	57
Manipulation of the tumour	13800	465
After resection	862	600
Autotransfusion	2772	58

hamic variables should be well controlled to maintain pulmonary blood flow.

It has been recommended that the circulating blood volume should be corrected with preoperative α adrenergic blockers, fluid replacement and β blockers if necessary.^{1,2,8} In a similar case, Tjeuw and Fong¹⁰ performed preoperative therapy with phenoxybenzamine and intravenous hydration and a Blalock-Taussig shunt was created before the operation for phaeochromocytoma, so that pulmonary blood flow from the systemic circulation would be maintained. In the present case no palliative shunt was performed before surgery, and the vasoconstricting effect of norepinephrine released from the tumour might contribute to maintain pulmonary blood flow, so that it may be dangerous to administer a vasodilator preoperatively because systemic vasodilation may upset the balance between systemic and pulmonary blood flow, and result in cardiovascular collapse. To perform careful vasodilatation and fluid loading preoperatively would be useful to prevent the wide swings in haemodynamic variables and pulmonary blood flow,² but volume loading is potentially undesirable for the patient with poor myocardial reserve.¹⁵ Thus we performed vasodilatation and haemodilution under continuous monitoring of arterial pressure, CVP, and SpO_2 just before the operation.

To manage the present case, real-time monitoring and immediate response to a change in patient's pulmonary blood flow were required. Although a pulmonary artery catheter is useful for measuring pulmonary blood flow in most situations, it was not used in the present case because of potential loss of indicator via the VSD, and because pulmonary stenosis would make the catheter flow measurement a useless determination. We expected that SpO_2 would be positively correlated with pulmonary blood flow and that a pulse oximeter would be a useful monitor. The SpO_2 correlated with heart rate better than with mean arterial pressure. Increased heart rate might shorten ventricular ejection time, and result in decreased pulmonary blood flow in the presence of pulmonary stenosis. Thus, control of heart rate was considered to be more important than that of blood pressure, and agents which

modify heart rate might be hazardous. In the previous report,¹⁰ SpO_2 was maintained above 90% throughout the operation and the decreases seen in the present case were not observed. This difference may have been related to the creation of a palliative shunt.¹⁰ Therefore, we suggest that a palliative shunt may be important for the anaesthetic management of similar cases, and creation of the shunt, if possible, should be considered before surgery for removal of the phaeochromocytoma.

Smith *et al.*¹⁶ reported that increases in blood pressure due to catecholamine administration are associated with autotransfusion. However, we did not observe haemodynamic changes during autotransfusion. This may have been because most of the blood was lost before tumour manipulation occurred, when the plasma concentrations of catecholamines were relatively low. In fact, norepinephrine and epinephrine concentration of the autotransfusate were $2772 \text{ pg} \cdot \text{ml}^{-1}$ and $58 \text{ pg} \cdot \text{ml}^{-1}$, respectively (Table). Furthermore, 400 ml of the depleted blood was infused over 60 min because there was no massive bleeding.

In conclusion, this case described the anaesthetic management of a patient with phaeochromocytoma associated with tricuspid atresia and pulmonary vascular stenosis. Pulmonary perfusion was heart-rate dependent. Pulse oximetry appeared to be a useful monitoring of pulmonary perfusion during anaesthesia in this patient.

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