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*Original Article*

# Anesthesia for encephalopyosis excision in two patients with single ventricle

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**Abstract:** Single ventricle is a rare complicated cyanosis congenital cardiovascular anomaly that accounts for only 2-3% of all congenital heart malformations. The single ventricular patient has one underdeveloped and one functional ventricule. The natural history of single ventricular patients is dismal, as most die before 1 year of age and only few survive to age 10 years. The anaesthetic management of single-ventricle patients for undergoing non-cardiac surgery remains a challenge. We present our anaesthetic management of encephalopyosis excision of two recent cases and discuss the rationale of using the chosen drug combination and the importance of adequate monitoring in selecting an anaesthetic technique based on the pathophysiology of the congenital cardiac lesion.

**Keywords:** Single ventricle, non-cardiac surgery, general anaesthesia, congenital heart disease

## Introduction

Recently two patients with single ventricle underwent encephalopyosis excision at our hospital. They were managed by a multidisciplinary team during their perioperative period. The encephalopyosis excision were uneventfully performed, Two patients under general anaesthesia and did well after surgery. We discuss the anaesthetic considerations in managing these high-risk patients.

## Case presentation

The first patient was a 14-year-old girl weight 35 kg, who complained of vomiting and fever for half a day, then was referred to our hospital. Preoperative diagnosis showed: (1) occipital lobe occupation in the right temporoparietal region, indicating a possibility of encephalopyosis; (2) complex congenital heart disease (cyanotic type), with single ventricle (left ventricular type), complete type endocardial cushion defect, pulmonary stenosis, arterial septal defect, and dexiocardia; (3) New York Heart Association class II~III. Physical examination revealed cyanosis and clubbing of her fingers. Clear breath sounds of bilateral lungs without rales or rhonchi, and audible 3/6 grade rumbling systolic murmur at 2-4 intercostal region of sternum's right border. Brain computed tomography showed occipital lobe lesion in the right temporoparietal region, suggesting a high probability of encephalopyosis. Chest X-rays revealed: (1) chronic pulmonary infection; (2) thickening of bilateral interlobar pleura; (3) situs inversus viscerum. Echocardiography indicated: (1) situs inversus, right atrial enlargement, a 21.0 mm discontinuity from the middle-lower segment of interatrial septum to decussation; (2) left ventricle as the main ventricular chamber with right ventricle being a small residual chamber, only a group of atrioventricular valves found to connect with the main ventricular chamber, and a 8.9 mm discontinuity from the upper segment of interventricular septum to decussation; (3) both the aorta and pulmonary artery originated from the main ventricular chamber, with the aortic valve located at the left anterior part and the pulmonary valve at the right posterior part (where the aorta and pulmonary artery travel along the left anterior and right posterior side, respectively) and normal aortic valve morphology and motion, accompanied with thickening, strengthened echo, limited opening and normal closure of the pulmonary valve; (4) CDFI indicated visible two-way shunt signals at the discontinuities of interatrial and interventricular

septa, accelerated fluid in systolic pulmonary orifice, visible moderate reflex at the atrial side of systolic atrioventricular orifice. Electrocardiography returned the following results: (1) sinus rhythm, HR of 89 beats/min; (2) left axis deviation (-30°); (3) I°AVB; (4) extreme clockwise translocation and dextrocardia. Blood routine examination revealed WBC at $15.33\times10^9$/L. Blood count returned basically normal results, with increased red blood cell, hemoglobin and hematocrit amounts, prolonged prothrombin time and partial thromboplastin time, as well as increased fibrinogen levels. The patient was submitted to resection of the right parietal-occipital occupation under general anesthesia.

The patient was placed in supine position, with cyanotic lips and nails, with basically normal respiration. Monitoring was carried out using ECG, $SpO_2$ and $P_{ET}CO_2$; peripheral venous pathways were opened, and left radial arterial cannulation was conducted to continuously monitor the arterial blood pressure, which showed BP 135/85 mmHg, HR 90 beats/min and $SpO_2$ 78%. Arterial blood gas analysis showed $FiO_2$ 21%, $PaO_2$ 44 mmHg, $PaCO_2$ 31 mmHg and $SaO_2$ 82%; $SpO_2$ was increased to 86% after mask oxygen-inspiration (2 L/min). Then, anesthesia induction was applied with 3 mg midazolam, gradual injection of sufentanil (each of 5 μg for a total of 20 μg), gradual injection of etomidate (each of 6 mg for a total of 18 mg), 4 mg vecuronium, and smooth endotracheal intubation. Afterward, the HR was 90 beats/min, BP 115/78 mmHg and $SpO_2$ 86%. Then, internal jugular vein catheterization was conduced to monitor central venous pressure (CVP). Anesthesia maintenance included 1.0 MAC sevoflurane and continuous infusion of 0.2-0.4 $\mu g.kg^{-1}.min^{-1}$ remifentanil. During operation, 3-8 $\mu g.kg^{-1}.min^{-1}$ dobutamine, 0.03-0.08 $\mu g.kg^{-1}.min^{-1}$ phenylephrine and 0.3-0.6 $\mu g.kg^{-1}.min^{-1}$ nitroglycerin were administered to maintain BP 100-130/55-78 mmHg, $SpO_2$ 85%-88%, HR 70-90 beats/min and CVP 15-17 $cmH_2O$. Arterial blood gas was tested intermittently during operation, and pH 7.36-7.40, $PCO_2$ 31-32 mmHg, $PaO_2$ 63-69 mmHg and lactate 0.6-1.0 were obtained. The surgery lasted 3 hours 40 minutes, with an intraoperative infusion of 1000 mL and urine volume of 1700 mL. After the operation, the patient was transferred to the ICU, where he woke up 4 hours later; examinations returned the following results: HR 75-85 beats/min, RR 15 beats/min, BP105-129/65-75 mmHg, $SpO_2$ 80-82% (inspired oxygen flow of 2 L/min), and a retracted endotracheal tube. The patient was transferred back to the neurosurgery ward the day after surgery, and discharged at day 13 postoperation.

The second patient was a 21-year-old man weight 45 kg. He complained of headache, dizziness and intermittent fatigue in his right extremities for more than 6 months. Preoperative diagnosis showed: (1) multiple occipital lobe occupations in the left temporoparietal region, indicating a high possibility of encephalopyosis; (2) congenital heart disease with single ventricle; (3) grade III cardiac function. His family declared that he had been admitted to our hospital several times due to heart failure. Physical examinations returned the following: chronic facies, clear mind, pigmentation of bilateral cheeks, lip cyanosis, failure to lie flat, rapid breathing, acropachy, rough breath sounds of bilateral lungs, audible 3/6 grade systolic murmur in 2-4 intercostal region of sternum's right border. Brain MRI revealed multiple occipital lobe occupations in the left temporoparietal region, suggesting a high probability of encephalopyosis. Chest X-rays revealed thickening or texture turbulence in bilateral lungs, and enlarged heart shadow. Echocardiography showed situs solitus, visible single ventricle and two groups of atrioventricular valves, left and right arrangement of aortic valve and pulmonary valve, with the aortic and pulmonary valves located at the left anterior and right posterior parts, respectively, both of which originated from a single ventricle, with normal inner diameters of aorta and pulmonary artery; the left and right pulmonary artery inner diameters were 11.6 mm and 12.3 mm, respectively. Electrocardiography yielded the following results: (1) sinus tachycardia, with HR at 109 beats/min; (2) right axis deviation (+105°); (3) $R/S_{V1}>1$; (4) left ventricular hypertrophy. Blood routine examination showed WBC at $13.38\times10^9$/L. Arterial blood gas analysis revealed $FiO_2$ of 21%, $PaO_2$ 27 mmHg, $PaCO_2$ 23 mmHg and $SaO_2$ 64%. Blood count returned basically normal results and unaltered clotting function. The patient underwent resection of the left temporoparietal-occipital occupation under general anesthesia.

The patient was in semi-recumbent position, with cyanotic lips and nails as well as rapid breathing. Monitoring was performed using

ECG, $SpO_2$ and $P_{ET}CO_2$; peripheral venous pathways were opened, and radial arterial cannulation was conducted to continuously monitor the arterial blood pressure, which showed BP 98/56 mm Hg, HR 90 beats/min and $SpO_2$ 70%. Arterial blood gas analysis showed $FiO_2$ 21%, $PaO_2$ 25 mmHg, $PaCO_2$ 33 mmHg and $SaO_2$ 64%, which was increased to 87% after mask oxygen-inspiration (5 L/min). Arterial blood gas showed $PaO_2$ 53 mmHg, $PaCO_2$ 29 mmHg and $SaO_2$ 89%. Anesthesia induction was carried out via sevoflurane inhalation with intravenous anesthetic drugs, gradual injection of sufentanil (each of 25 μg for a total of 100 μg), 10 mg etomidate, 4.5 mg vecuronium, and smooth endotracheal intubation, after which HR was 90 beats/min and BP 89/50 mmHg. Then, subclavian vein catheterization was conducted to monitor CVP. Anesthesia maintenance included 1.0 MAC sevoflurane, intermittent injection of vecuronium and fentanyl. During the operation, 8-15 $\mu g.kg^{-1}.min^{-1}$ dopamine was applied to maintain BP 75-100/40-60 mmHg, $SpO_2$ 89%-95%, HR 90-120 beats/min and CVP 4-6 $cmH_2O$. Arterial blood gas was tested intermittently during the operation, and pH 7.36-7.40, $PCO_2$ 29-40 mmHg, $PaO_2$ 53-137 mmHg, lactate 1.6-1.9 and normal electrolyte levels were obtained. The operation lasted 3 hours 15 minutes, for an intraoperative infusion of 3650 mL and urine amount of 3650 mL. After surgery, the patient was transferred to the ICU, where examinations on operation day returned HR 68 beats/min, RR 18 beats/min, BP 106/64 mmHg and $SpO_2$ 89%; meanwhile, examinations the day after surgery showed HR 67 beats/min, RR 20 beats/min, BP 105/63 mmHg and $SpO_2$ 88%. The patient was transferred back to the neurosurgery ward at day 3 post-operation, and discharged at day 15 after surgery.

## Conclusions

Single ventricle is a rare complex cyanotic congenital cardiac malformation, accounting for 1~2% of all congenital cardiac malformations. The definition of single ventricle has been controversial since Holmes reported a heart with a single ventricle in 1924. Currently, according to the Classification and nomenclature of congenital heart surgery, single ventricle is considered when the atrium is only connected with a well-developed dominant ventricle [1]. Furthermore, single ventricle classification is also inconsistent: it can be subtyped according to the anatomic characteristics of ventricle, or based on aortic arrangement. However, regardless of subtyping, the major anatomic characteristic of single ventricle is that only a group of common atrioventricular valves or two groups of atrioventricular valves are connected to the functional ventricle, in order to import the atrioventricular blood into the functional ventricle [2]. The two cases presented here were diagnosed as single ventricle according to anatomic characteristics. Patients with single ventricle often have other cardiac malformations. While case 1 was accompanied by pulmonary stenosis, atrial septal defect and dextrocardia, case 2 also showed transposition of great arteries.

The pathophysiological changes in these patients depend on the development of the cardiac malformations, pulmonary valve and vascular bed, size of total pulmonary resistance, and functional status of the ventricle. The common pathophysiological characteristics include the following. (1) Since the single ventricle accepts blood from mitral and tricuspid valves, the pulmonary venous blood (arterial blood) and systemic blood (venous blood) tend to be thoroughly mixed in the ventricle, leading to similar oxygen saturation levels in the aorta and pulmonary artery. (2) Since the aorta and pulmonary artery originate from the same cardiac chamber, the ventricular output is the sum of pulmonary flow quantity (Qp) and systemic flow quantity (Qs). Therefore, single ventricle belongs to the balanced shunts in the classification of congenital heart diseases [3], whose characteristic is that right and left ventricular ejections directly enter the pulmonary and systemic circulations, where flow quantities are dependent entirely on the pulmonary vascular resistance (PVR) and systemic vascular resistance (SVR). For the two cases described above, pathophysiological changes differed since they were accompanied by different types of cardiac malformation. Case 1 had significant pulmonary stenosis, leading to reduced blood flow into the pulmonary circulation and therefore significant cyanosis as well as increased red blood cell numbers over time. In case 2, bilateral pulmonary arteries were developed normally, showing increased pulmonary blood flow. However, with the increased ventricular load and cyanosis appearance, the ventricular

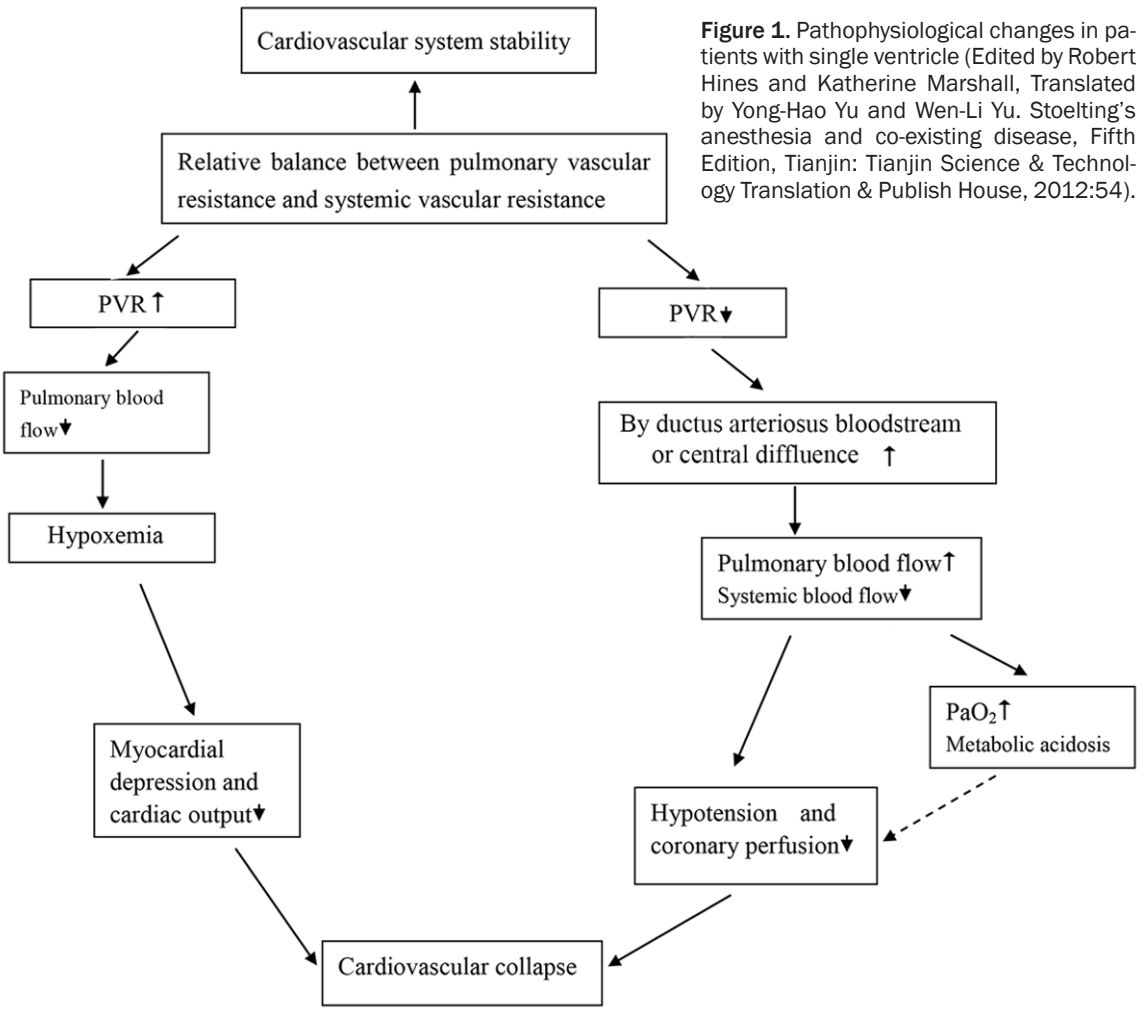


**Figure 1.** Pathophysiological changes in patients with single ventricle (Edited by Robert Hines and Katherine Marshall, Translated by Yong-Hao Yu and Wen-Li Yu. Stoelting's anesthesia and co-existing disease, Fifth Edition, Tianjin: Tianjin Science & Technology Translation & Publish House, 2012:54).

function was further impaired, commonly resulting in congestive heart failure. The pathophysiological changes in patients with single ventricle are summarized in **Figure 1**.

For the two patients, anesthesia management aimed to achieve two common targets, i.e. to maintain (1) cardiovascular function stability and (2) the PVR and SVR balance, so as to optimize blood flow, ensuring sufficient oxygenation and systemic perfusion. However, the difference in the treatment of the two patients mainly concerned pulmonary blood decrease prevention (case 1), and pulmonary blood increase prevention (case 2).

Measures for assuring cardiovascular function stability include: hemodynamic stability maintenance, preventing serious changes of cardiac function as well as balancing systemic and pulmonary circulations. At present, most studies suggested that systemic circulation tends to provide the highest oxygen supply when Qp/Qs ratio is equals to or close to 1/1 by balanced circulation [4], while systemic circulation is likely to severely decline when Qp/Qs is larger or lower than 1/1. Therefore, it is more ideal to maintain a $SaO_2$ value of 75%~80% [5, 6]. However, recent studies have shown that systemic circulation might show severe oxygen supply shortage even with $SaO_2$ strictly controlled to a range of 75%~80% [7]; in addition, oxygen saturation alone was shown to be a poor guide for estimating Qp/Qs [8]. Thus, the following measures were adopted in this study: (1) Anesthesia induction and maintenance using venous anesthetics (midazolam, etomidate and fentanyl) and inhalation of sevoflurane were selected, which resulted in smaller impact on cardiovascular functions, preventing myocardial depression. (2) Invasive arterial pressure and CVP were closely monitored with

blood volume timely replenished. Blood pressure was more ideally maintained in case 1 (100-130/55-78 mmHg) as well as HR (70-90 beats/min). Meanwhile, blood pressure was lower with larger fluctuations in case 2 due to poor cardiac function: it was maintained at BP 75-100/40~60 mmHg, HR 90-120 beats/min, with a lower CVP. (3) Hypotensive patients without hypovolemia would be given positive inotropic drugs (e.g. dopamine and dobutamine) as early as possible.

Maintaining the balance between PVR and SVR, and optimizing blood flow serve to ensure adequate oxygenation and systemic perfusion. Since methods to directly monitor PVR and SVR are scarce, the following measures were adopted in this study: (1) $SpO_2$ was monitored to reflect oxygenation, maintained at 85%-88% in case 1, and 89%-95% in case 2, both showing reasonable oxygenation. (2) The pH value, lactate and urine amounts in arterial blood were monitored to reflect the systemic perfusion. In case 1, pH, lactate level and urine amount were 7.40-7.44, 0.6-1.0 and 1450 mL (anesthesia duration was 5 hours and 40 minutes), respectively, with 7.36-7.47, 1.6-1.9 and 3050 mL (anesthesia duration was 7 hours), respectively, in case 2, suggesting that both cases had good systemic perfusion.

In order to prevent pulmonary blood reduction in case 1, the following measures were adopted: (1) Increased SVR resistance to enhance pulmonary blood flow and ensure oxygenation. Therefore, a small dose of vasoactive drug, the α-receptor agonist phenylephrine hydrochloride, was adopted to increase systemic resistance. (2) Moderate hypocapnia was maintained ($PaCO_2$ maintained at 31-32 mmHg) to increase inspired oxygen concentration, hereby maintaining normal functional residual capacity and preventing acidosis. In order to prevent pulmonary blood increase in case 2, the following measures were applied: normal or slightly elevated $PaCO_2$, application of PEEP, restrictions of inspired oxygen concentration, and increased PVR could be applied to decrease pulmonary blood flow, which was conducive to systemic perfusion. However, case 2 treatment had the following deficiencies: (1) $PaCO_2$ was maintained at a lower level of 29-40 mmHg; (2) PEEP was not applied; (3) 100% pure oxygen was inhaled due to limited conditions (lacked of oxygen-air mixer); (4) $SpO_2$ was maintained at 89%-95% (such excessive pulmonary blood flow must be obtained by reducing systemic blood flow). Therefore, increasing pulmonary blood flow in such patients might aggravate congestive heart failure and cause pulmonary steal, leading to hypotension, reduced coronary perfusion, hypoperfusion of major organs and metabolic acidosis. In this patient, blood pressure remained lower after administration of a large dose of dopamine; this might be associated with reduced systemic blood flow (also associated with hypovolemia). The patient was fortunate not to suffer severe complications such as acute congestive heart failure, arrhythmias, myocardial ischemia, and failure of major organs. In addition, bubbles must be removed from intravenous perfusion pathways to prevent aeremia.

Although these two patients both suffered from single ventricle, they received different treatments due to distinct pathophysiology. Therefore, in order to conduct non-cardiac surgery in patients with complex heart diseases, understanding their pathophysiological features is more important than selecting anesthetic drugs.

## Acknowledgements

All subjects were Han Chinese, and written informed consent was obtained from parents of all the subjects before participating in the clinical trial.

## Disclosure of conflict of interest

None.

## Authors' contribution

Kaiyun Fang, Fujuan He and Fangxiang Zhang carried out the studies, participated in collecting data, and drafted the manuscript. Suixiang Yin and Kedong Jiang participated in its design. Kaiyun Fang and Fujuan He helped to draft the manuscript. All authors read and approved the final manuscript.

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