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# 17 Monitored anesthesia care for craniotomy in a patient with Eisenmenger syndrome: A case report

Ri HS *et al.* MAC in Eisenmenger syndrome

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## Abstract

### BACKGROUND

Eisenmenger syndrome (ES) is an uncorrected congenital cardiac defect with a left-to-right shunt, leading to pulmonary arterial hypertension. Patients with ES are susceptible to hemodynamic alterations during noncardiac surgery with general anesthesia, which increases perioperative morbidity and mortality. Monitored anesthesia care (MAC) is often used during minor procedures in patients with cardiac disease. However, few reports on MAC in patients with ES exist.

### 8 CASE SUMMARY

1 A 49-year-old man was admitted for a severe headache lasting 30 d. He had been diagnosed with a large perimembranous ventricular septal defect (VSD) with bidirectional shunt flow and pulmonary arterial hypertension 10 years ago. A round mass in the right frontal lobe was revealed by Magnetic resonance imaging. Stereotactic aspiration using a neuronavigation system was performed under MAC. The patient was stayed in the hospital for 5 d, and discharged without complications.

### CONCLUSION

MAC may be effective for craniotomy in patients with ES.

**Key Words:** Case report; Eisenmenger syndrome; Pulmonary hypertension; Sedation; Anesthesia; Craniotomy; Case report

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**Core Tip:** Monitored anesthesia care (MAC) is often used during minor procedures in patients with cardiac disease. However, there are few reports on MAC in patients with Eisenmenger syndrome (ES). We report a case of MAC using dexmedetomidine and remifentanyl for craniotomy in a patient with ES.

## INTRODUCTION

Eisenmenger syndrome (ES) is an advanced form of pulmonary arterial hypertension caused by congenital heart disease<sup>[1]</sup>. ES is characterized by irreversible severe pulmonary hypertension and a left-to-right shunt. When pulmonary artery pressure reaches the systemic level, ES cannot be resolved<sup>[1]</sup>. As the life expectancy of patients with ES has improved in recent decades, the number of adult patients undergoing noncardiac surgery has steadily increased<sup>[2,3]</sup>. Patients with ES undergoing noncardiac surgery are at high risk for fatal or life-threatening complications during the perioperative period<sup>[4-8]</sup>; perioperative systemic inflammatory response, hypoxemia, or hypercapnia and activation of the sympathetic nervous system can lead to deterioration of pulmonary hypertension and right heart failure in patients with ES<sup>[9-11]</sup>. Therefore, anesthetic management of patients with ES is challenging, and comprehensive, evidence-based guidelines for anesthetic management are needed<sup>[12]</sup>. Monitored anesthesia care (MAC) is an anesthetic technique that provides conscious sedation with a local anesthetic and sedative for minor surgeries, including cardiovascular interventions and craniotomy. Herein, we report the case of a patient with ES who underwent MAC for craniotomy for a brain abscess.

## CASE PRESENTATION

### Chief complaints

A 49-year-old man (170 cm, 48.4 kg) was admitted for a severe headache.

### *History of present illness*

The headache lasted for 30 d.

### *History of past illness*

He had been <sup>1</sup>diagnosed with a large perimembranous <sup>1</sup>ventricular septal defect (VSD) with bidirectional shunt flow and pulmonary arterial hypertension 10 years ago. He had exertional dyspnea and heart function classified as <sup>1</sup>New York Heart Association Class III and was on medications, including bosentan, furosemide, and spironolactone.

### *Personal and family history*

He was on medications, including bosentan, furosemide, and spironolactone.

### *Physical examination*

He had exertional dyspnea and heart function <sup>3</sup>classified as New York Heart Association Class III

### *Laboratory examinations*

Preoperative blood laboratory assessments, such as complete blood count and liver and kidney tests, were unremarkable.

### *Imaging examinations*

Electrocardiography (ECG) revealed sinus rhythm with left ventricular hypertrophy. Cardiac catheterization demonstrated a pulmonary artery pressure of 101/62 mmHg, pulmonary vascular resistance of 15.1 Woods units, and cardiac output of 3.44 L/min. Echocardiography revealed a large VSD (18 mm) with bidirectional <sup>11</sup>shunt flow, a D-shaped left ventricle with normal systolic ejection function (56%), <sup>11</sup>right ventricular hypertrophy with preserved <sup>11</sup>right ventricular contractibility, and resting pulmonary hypertension with trivial <sup>11</sup>tricuspid regurgitation. Magnetic resonance imaging revealed

a round mass in the right frontal lobe, which was suspected to be a brain abscess (Figure 1).

### **FINAL DIAGNOSIS**

We suspected that the patient had a brain abscess in the area of the right frontal lobe.

### **TREATMENT**

The patient was scheduled to undergo stereotactic aspiration using a neuronavigation system (StealthStation® S7, Medtronic, Inc., Louisville, CO, United States) under MAC. Premedication was not administered. Standard monitoring included ECG, pulse oximetry, invasive blood pressure measurement, and esophageal stethoscope temperature measurement.

Before induction of MAC, a 20 G catheter was inserted into the left radial artery, and arterial blood gas analysis on room air showed a pH of 7.49, partial pressure of carbon dioxide (PaCO<sub>2</sub>) of 30 mmHg, partial pressure of oxygen (PaO<sub>2</sub>) of 53 mmHg, bicarbonate ion (HCO<sub>3</sub><sup>-</sup>) concentration of 22.1 mmol/L, and oxygen saturation (SaO<sub>2</sub>) of 89.9%. Cardiac output (CO), stroke volume (SV), and stroke volume variation (SVV) were continuously measured using a FloTrac®/Vigileo device (Edwards Lifesciences, Irvine, CA, United States). CO, SV, and SVV were 6.1 L/min, 100 mL/beat, and 5%, respectively. Oxygen was provided at 20 L/min with a fraction of inspired oxygen (FiO<sub>2</sub>) of 0.95 using a high-flow nasal cannula (HFNC) (Flo Easy™, Westmed, Inc., Tucson, AZ, United States) during the perioperative period. MAC was induced with dexmedetomidine at a loading dose of 1 µg/kg for 30 min. After infiltration of 2% lidocaine into the insertion site, a central venous catheter was inserted into the right internal jugular vein. During the procedure, remifentanyl and dexmedetomidine were infused at the rate of 0.02 µg/kg/min and 0.8-1.2 µg/kg/h, respectively. To manage the pulmonary hypertension, remodulin was infused intravenously at a rate of 0.01 µg/kg/min. The patient was placed in the supine position on a horseshoe headrest. Local anesthesia at the incision site was induced with 0.5% bupivacaine with 1:200000

epinephrine, and craniotomy was performed. Stereotactic aspiration was performed, and 20 mL of yellow discharge was extracted. During surgery, the heart rate was 60-80 beats/min, spot oxygen saturation was 97%-99%, mean arterial pressure was 99-86 mmHg, and body temperature was 36.6 °C. CO, SV, and SVV were 4.6-6.1 L/min, 69-100 mL/beat, and 5%-9%, respectively. Arterial blood gas analysis showed that pH, PaCO<sub>2</sub>, PaO<sub>2</sub>, HCO<sub>3</sub><sup>-</sup>, and SaO<sub>2</sub> were 7.42, 42.0 mmHg, 94.0 mmHg, 27.2 mmol/L, and 97.0%, respectively. The surgery lasted for 60 min, and the patient tolerated the surgery without dysrhythmia or hemodynamic changes. After discontinuation of MAC, the patient was conscious and transferred to the surgical intensive care unit (ICU).

In the surgical ICU, oxygen was provided at a rate of 4 L/min via a facemask.

## OUTCOME AND FOLLOW-UP

The patient cooperated well. The patient was transferred to the ward on postoperative day 3, and 2 d later, he was discharged without complications. The timeline of this case is shown in Table 1.

## DISCUSSION

ES is defined as an untreated congenital cardiac defect with a left-to-right shunt that results in elevation of pulmonary arterial pressure at a systemic level<sup>[13]</sup>. The main anesthetic goals in patients with ES are as follows: (1) Maintain euvolemia; (2) optimize systematic vascular resistance; and (3) avoid the elements that increase pulmonary vascular resistance (pain, hypoxia, and hypercapnia)<sup>[3,14,15]</sup>.

MAC with sedation has been implemented in anesthesia management of patients with ES, even for minor surgical procedures. The major concerns in MAC are (1) hypoxia and hypercarbia due to oversedation and lack of adequate airway support, and (2) potential agitation and uncontrollable behaviors due to the paradoxical reaction of sedative agents<sup>[13]</sup>.

Dexmedetomidine, an alpha-2 adrenoreceptor agonist, has sedative and analgesic effects. It can induce sedation without respiratory depression and could, therefore, be a



good choice to sedate patients with Eisenmenger features, while avoiding hypoxemia or hypercapnia. Intravenous anesthetics, such as propofol, have myocardial depression and respiratory inhibition effects, which can cause problems in patients with severe pulmonary arterial hypertension. Dexmedetomidine is less likely to cause paradoxical reactions than other anesthetics, such as benzodiazepines. The disadvantage of dexmedetomidine is the slow time of action. However, neuronavigation preparation took 30 min; therefore, it was used without inconvenience.

Although the use of opioids can cause respiratory depression and chest wall rigidity, we used a minimal dose of remifentanyl while providing oxygen through an HFNC and adjusting it as needed. This was because intraoperative pain could not be sufficiently controlled with dexmedetomidine alone. HFNC is a recent noninvasive technique that provides heated and humidified gas. HFNC was applied to patients with pulmonary arterial hypertension to avoid hypoxia and hypercarbia, thereby decreasing the need for intubation<sup>[16]</sup>. In this case, HFNC with 0.95% FiO<sub>2</sub> was performed, and the intraoperative vital signs were stable.

FloTrac<sup>®</sup>/Vigileo is a minimally invasive hemodynamic monitoring device that can be used via the radial artery<sup>[17]</sup>. Advanced hemodynamic variables (CO, SV, and SVV) were monitored, but systemic vascular resistance was not. Monitoring of advanced hemodynamic variables allowed the patient to remain stable without the use of vasoactive agents. A pulmonary artery catheter could be used; however, it is difficult to place it appropriately in patients with pulmonary hypertension without sedation and poses a risk of fatal complications.

## CONCLUSION

MAC with dexmedetomidine and remifentanyl may be effective for craniotomy in patients with ES. Supplementing oxygen *via* HFNC is advisable to avoid hypoxia.

## **Figure Legends**

**Figure 1 Magnetic resonance imaging finding.** Magnetic resonance imaging revealed a round mass in the right frontal lobe, which was suspected to be a brain abscess.



**Table 1 Case report timeline**

<b>Item</b>		<b>Timeline</b>
Preoperative	1	Admission for a severe headache lasting 30 d
	2	Past history-Large perimembranous VSD with bidirectional shunt flow and pulmonary arterial hypertension 10 years ago
	3	MRI-A round mass in the right frontal lobe
	4	Plan-craniotomy under MAC
Perioperative	5	Arterial cannulation and arterial blood gas analysis
	6	Advanced hemodynamic monitoring-CO, SV, SVV
	7	Oxygen supply-high-flow nasal cannula
	8	MAC induction with dexmedetomidine loading
	9	Central venous catheterization
	10	Remifentanil, remodulin infusion
	11	Neuronavigation system set
	12	Craniotomy and stereotactic aspiration
Postoperative	13	ICU for 3 d
	14	Discharge on POD 5

MRI: Magnetic resonance imaging; MAC: Monitored anesthesia care; CO: Cardiac output; <sup>14</sup>SV: Stroke volume; SVV: Stroke volume variation; ICU: Intensive care unit; POD: Postoperative day; VSD: Ventricular septal defect.

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