Transient Cardiac Arrest in Patient With Left Ventricular Noncompaction (Spongiform Cardiomyopathy)

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Left ventricular noncompaction (LVNC), also known as spongiform cardiomyopathy, is a severe disease that has not previously been discussed with respect to general anesthesia. We treated a child with LVNC who experienced cardiac arrest. Dental treatment under general anesthesia was scheduled because the patient had a risk of endocarditis due to dental caries along with a history of being uncooperative for dental care. During sevoflurane induction, severe hypotension and laryngospasm resulted in cardiac arrest. Basic life support (cardiopulmonary resuscitation) was initiated to resuscitate the child, and his cardiorespiratory condition improved. Thereafter, an opioid-based anesthetic was performed, and recovery was smooth. In LVNC, opioid-based anesthesia is suggested to avoid the significant cardiac suppression seen with a volatile anesthetic, once intravenous access is established. Additionally, all operating room staff should master Advanced Cardiac Life Support/Pediatric Advanced Life Support (including intraosseous access), and more than 1 anesthesiologist should be present to induce general anesthesia, if possible, for this high-risk patient.

Key Words: Left ventricular noncompaction; Spongiform cardiomyopathy; General anesthesia; Laryngospasm; Cardiac arrest; Dental treatment; Heart disease.

Left ventricular noncompaction (LVNC), also known as spongiform cardiomyopathy, is a recently described, rare, congenital cardiomyopathy with a poor long-term prognosis. It is characterized by multiple prominent trabeculations with deep intertrabecular recesses within the ventricular walls. The disease usually affects the left ventricle with severe arrhythmias, systemic emboli, eventual heart failure, and sudden death.¹⁻⁷ Although this disease is important, no case reports have described general anesthesia in patients with LVNC. We experienced a case of transient cardiac arrest in a child with LVNC during induction of general anesthesia for a dental procedure.

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CASE REPORT

The patient was a 4 year, 2 month old boy (height 97 cm, weight 12 kg). He had carious teeth with odontogenic infection and the potential for infective endocarditis and further deterioration of his health. However, safe dental treatment without general anesthesia was impossible because he was uncooperative. The patient came to our hospital after referral from his pediatrician. He was diagnosed with an atrial septal defect at 1 month of age. He was diagnosed with LVNC and chronic heart failure at 4 months. The patient was taking alpha- and beta-blockers, an angiotensin-converting enzyme (ACE) inhibitor, digitalis, and diuretics. He was sedentary in daily life. Climbing stairs, running, or crying resulted in breathlessness and cyanosis within approximately 1 minute. We evaluated him as American Society of Anesthesiologists (ASA) physical status 3. Preanesthetic blood tests were difficult to obtain because of his uncooperativeness and needle

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Anesthesia record.

phobia. Even his pediatrician performed only absolutely necessary tests to avoid anoxic episodes. A pre-anesthetic echocardiogram revealed a left ventricular ejection fraction of 54 to 70%. No abnormality in valve function, pulmonary pressure, middle septum, or posterior wall of the left ventricle was observed. Although the cardiothoracic ratio on chest x-ray was 57% 1 year previously, it had improved to 50% by the time of our preanesthetic examination. Heart rate and pulse oximetry (SpO₂) were 100 beats per minute (bpm) and 98%, respectively, at rest on room air. The family consented to dental treatment to be provided under general anesthesia because they recognized the danger of not treating the infection and were concerned about using restraint to accomplish dental treatment without general anesthesia. They were informed of the risks of general anesthesia before providing consent. We planned an opioid-based anesthetic immediately after insertion of the intravenous catheter during deep sedation by inhalation anesthesia. In addition, intraoperative continuous administration of olprinone, a phosphodiesterase inhibitor in the same class as milrinone, was planned to maintain the patient's heart function.

The patient had nothing to eat for 10 hours and nothing to drink for 3 hours before the procedure. Normal medications, except for alpha- and beta-blockers, were continued and were taken at 10 and 3 hours before operation. Preanesthetic blood pressure (BP),

heart rate (HR), SpO₂, and body temperature were 100/70 mm Hg, 96 bpm, 98%, and 36.1°C, respectively.

At 9:35 AM, the patient was escorted into the operating room with his mother. This was done without premedication. The anesthesia record is shown in the Figure. He quietly accepted the mask and inhaled a mixture of sevoflurane with oxygen (O₂)/nitrous oxide (N₂O). After the patient was sedated, his mother left the operating room, and assisted ventilation was begun. However, initial measurement of BP after assisted ventilation was already too low to measure. We decreased the sevoflurane concentration immediately and tried to quickly insert the intravenous catheter. Then laryngospasm occurred owing to stimulation under light anesthesia. It became impossible to ventilate the patient and SpO₂ decreased sharply, even with positive-pressure 100% O₂.

Endotracheal intubation was also impossible owing to spasm of the vocal cords. Thereafter, an intravenous catheter was inserted in an antecubital vein, and 2 mg vecuronium and 0.025mg fentanyl were administered immediately. At this time, there was no reading on the monitor for SpO₂ or BP, and we could not palpate a carotid pulse. Very quickly, the electrocardiogram (ECG) demonstrated severe bradycardia. A nurse started cardiopulmonary resuscitation (CPR) immediately. Approximately 1 minute after CPR had been started, ventilation became possible, but only gradually. Ac-

cordingly, the values of SpO₂, HR, and BP slowly improved. Soon afterward, endotracheal intubation could be performed. No remarkable abnormality such as metabolic acidosis or hypoxia was noted in an arterial blood gas analysis done immediately after the patient was stabilized. We decided to continue the anesthesia and the dental procedure because the patient still had the potential for infective endocarditis due to dental caries and untreated odontogenic infection. Respiratory and circulatory functions were stable under fentanyl and low-concentration sevoflurane anesthesia with continuous administration of olprinone. Awakening and recovery were smooth after the procedure, and the patient was discharged from the hospital the next morning without sequelae.

DISCUSSION

LVNC and Barth syndrome are recently described rare congenital cardiomyopathies caused by multiple structural cardiac abnormalities.^{1–7} LVNC is characterized by multiple prominent trabeculations with deep intertrabecular recesses in ventricular walls.^{1–7} Mutation of a novel gene in G4.5 has been reported in these individuals.^{1,2} Although LVNC is technically an unclassified cardiomyopathy, classification as a specific cardiomyopathy seems to be more appropriate.³

In Ozkutlu's paper, the mean age was 3.5 years in the 12 patients with LVNC that were studied.⁴ However, several reports have described adult patients.³ The diagnosis is based on findings on two-dimensional echocardiography; currently magnetic resonance imaging (MRI) is used more often.^{5,7} This disease involves left ventricular dysfunction, heart failure, severe arrhythmias, systemic embolism, pulmonary stenosis, coarctation of aorta with aberrant origin of the right subclavian artery, ventricular septal defect, partial anomalous pulmonary venous return, and sudden death.^{4,5,7} Observed rhythm abnormalities included Wolff-Parkinson-White syndrome, supraventricular tachycardia, ventricular extrasystoles, left bundle branch block, and complete atrioventricular block.^{4,6}

In a retrospective study of long-term follow-up of 34 adults with LVNC, major complications consisted of heart failure (53%), thromboembolic event (24%), ventricular tachycardia (41%), death (35%), sudden death (18%), death by end-stage heart failure (12%), and death by other cause (6%).³ Treatment of this disease included heart transplantation (12%) and implantation of automated cardioverter/defibrillators (12%).³ Other reports describing the risk stratification include heart failure therapy and oral anticoagulation.³ Therefore, LVNC is a very important disease in general anesthesia management.

No reports have documented general anesthesia for patients with LVNC. We subsequently referred a patient with dilated cardiomyopathy (DCM) to the anesthesia department because the pathogenesis of both diseases is similar from an anesthetic management standpoint. Therefore, severe hypotension due to volatile anesthesia administration could be predicted. In fact, cardiac function is suppressed remarkably by volatile anesthesia in patients with LVNC. Thus, intravenous induction of general anesthesia is recommended for these patients with maintenance of anesthesia utilizing an opioid-based technique. A child with heart disease who refuses venipuncture and has an intravenous catheter placed under restraint may induce high-risk accidents (anoxic spell, acute heart failure, malignant syndrome, embolism, shock, asphyxia, aspiration, etc.) as the result of crying and lack of cooperation.

Controversy continues regarding whether it is necessary to secure intravenous (IV) access prior to induction of general anesthesia. This may be an issue for discussion among anesthesiologists. In fact, crying and exertion-induced breathlessness and cyanosis in this patient when visiting the patient's pediatrician also suggested that anoxic spells may occur if restraint were used. Although a sedative (intramuscular, oral, or suppository) as a premedication might be effective enough to allow insertion of the IV catheter in such a patient, unexpected cardiac suppression or excitation might be induced owing to an inappropriate dose. This patient also refused even oral premedication with syrup or juice. Therefore, we decided to escort the patient to the operating room with his mother instead of attempting premedication, and we then induced deep sedation by inhalation anesthesia.

We were unable to detect the patient's carotid pulse when severe bradycardia was recognized on the ECG. We diagnosed pulseless electrical activity (PEA) based on the Advanced Cardiac Life Support (ACLS)/Pediatric Advanced Life Support (PALS) algorithms of the American Heart Association (AHA).^{8,9} PEA is a category of cardiac arrest in which there may be a normal electrical pattern of the heart, but not enough cardiac output to produce a pulse. Fortunately, not only the anesthesiologists but also the operating room nurses had taken ACLS/PALS previously. In fact, the nurse correctly began CPR based on AHA guidelines. It was speculated that the chest compressions circulated both vecuronium and fentanyl. Additionally, had we not been able to quickly establish venous access, intraosseous infusion techniques would have been very useful during this case. We strongly suggest that not only anesthesiologists but also nurses need to be current with AHA guidelines, even though these emergencies are rare. Although the responsible anesthesiologist felt real panic, his supervisor directed the code and responded properly. We suggest that for difficult, high-

risk cases such as the one described, for the patient's safety, 2 anesthesiologists should be present if possible to induce general anesthesia.

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