Pronounced QT Prolongation During General Anesthesia in a Child with Left Ventricular Noncompaction Cardiomyopathy: A Case Report

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We report the case of an 8-year-old boy with left ventricular noncompaction cardiomyopathy (LVNC) and QT prolongation who experienced further prolongation of the QTc during general anesthesia for extraction of a maxillary mesiodens. Pronounced prolongation of the QTc was observed after induction of general anesthesia with thiamylal and during emergence. No notable fluctuations in blood pressure, heart rate, and estimated continuous cardiac output were observed. We considered it likely that the QT prolongation was triggered by thiamylal and increased sympathetic nervous system activity. During general anesthesia for children with LVNC and QT prolongation, it is necessary to monitor intraoperative hemodynamic fluctuations and prepare for the possible occurrence of arrhythmias.

Key Words: Left ventricular noncompaction; General anesthesia; QTc; QT prolongation; Cardiac output.

Left ventricular noncompaction cardiomyopathy (LVNC) is a very rare congenital cardiac muscle disorder in which the normal condensation or compaction of the myocardium is impaired.[1] The resulting myocardium appears thick and spongy rather than developing as smooth and firm. Although presentation can vary, LVNC can cause heart failure similar to dilated cardiomyopathy, embolism as a result of mural thrombosis, or electrocardiographic abnormalities including fatal arrhythmias, and it can be associated with neuromuscular diseases.[2]

CASE PRESENTATION

An 8-year-old boy (height, 137.3 cm; weight, 36.4 kg; body mass index, 19.3 kg/m^2) diagnosed as having LVNC was scheduled to undergo general anesthesia for the extraction of a maxillary mesiodens. The patient had undergone general anesthesia for cardiac catheterization 2 years prior, had no other notable medical comorbidities or allergies, and was taking bisoprolol, which he had been on for 2 years. His preoperative chest radiograph revealed a cardiothoracic ratio of 50%, and his preoperative laboratory data showed a slightly elevated brain natriuretic hormone level (47 pg/mL). An electrocardiogram obtained 1 month before surgery demonstrated prolongation of the corrected QT interval (QTc) of 529 ms using the Bazett formula (QTc = QT time/$\sqrt{}$RR interval) with flat or inverted T waves in all leads. He was suspected of having long-QT syndrome (LQTS) according to the diagnostic criteria (score: 3) of Schwartz and Crotti.[3] No major genetic abnormalities associated with LVNC were detected in his family, and the cause of the QT prolongation in this patient was not clear. However, it was reported that a *SCN5A* gene mutation in patients with LVNC could be involved in arrhythmias such as QT prolongation.[4]

On the day of surgery, standard anesthesia monitors were applied that included electrocardiography, nonin-

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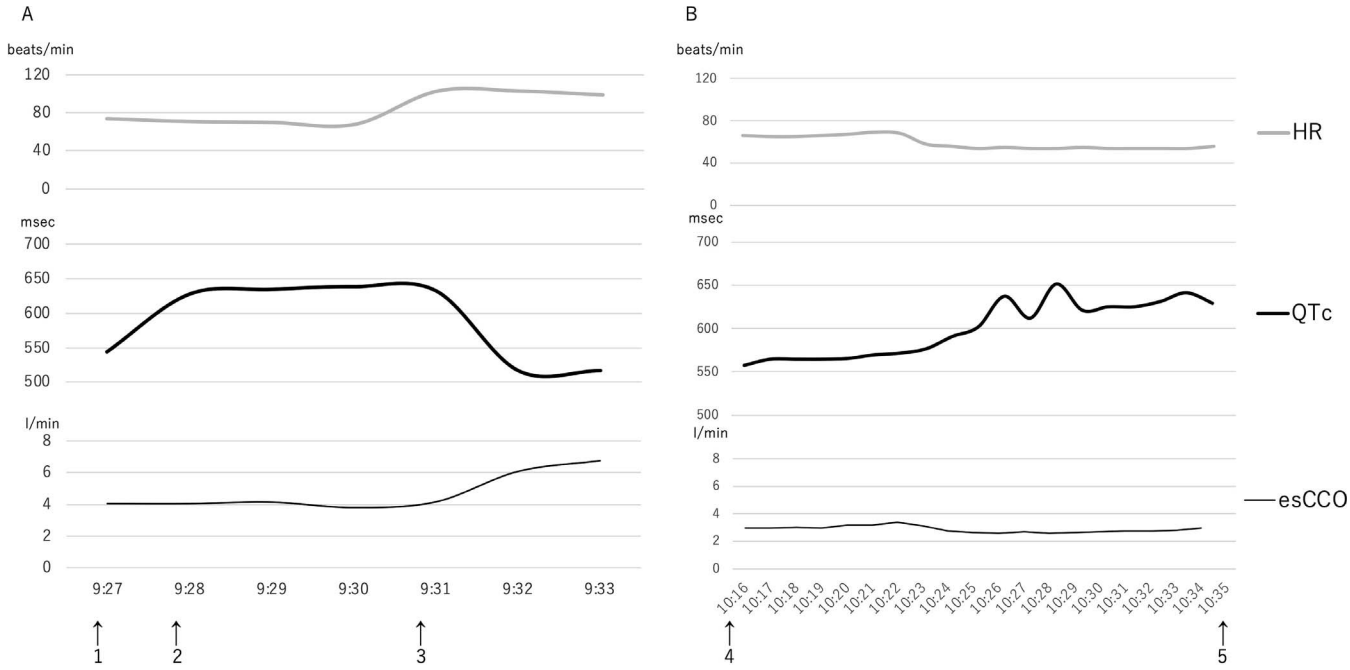
Figure. QTc Prolongation After IV Administration of Thiamylal (A) and During Emergence (B).



Caption: (A) 1: One minute after IV administration of thiamylal (150 mg); 2: 2 minutes after IV administration of thiamylal (150 mg); 3: end of intubation. (B) 4: End of the operation; 5: extubation.
Abbreviations: IV, intravenous; HR, heart rate; QTc, corrected QT; esCCO, estimated continuous cardiac output.

vasive blood pressure (NIBP), pulse oximetry, a peripheral nerve stimulator to assess neuromuscular blockade depth, and capnography. During general anesthesia, QTc, estimated continuous cardiac output (esCCO; Nihon Kohden; measured from pulse transit time using electrocardiography electrodes), esCCO compatible pulse oximetry, and NIBP were also measured, and a defibrillator was prepared in the operating room because QT prolongation has been associated with life-threatening arrythmias like torsade de pointes.

After inserting a 22-gauge cannula into a vein of the left forearm, the patient was induced using remifentanil (0.2 μg/kg/min), thiamylal (150 mg), and rocuronium (30 mg), and general anesthesia was maintained with sevoflurane (2%), air (2 L/min), oxygen (1 L/min), and remifentanil (0.1–0.2 μg/kg/min). Local infiltration anesthesia was performed using 1.8 mL of 2% lidocaine with 1:73,000 epinephrine immediately before and at the end of the operation (total 72 mg lidocaine, 0.049 mg epinephrine). The patient's QTc before induction of general anesthesia was 452 ms. The QTc after intravenous administration of thiamylal (150 mg) and during intubation were 627 and 638 ms, respectively. However, the patient's QTc shortened after intubation (Figure A). The QTc was 530 to 580 ms during maintenance of general anesthesia, although it became prolonged to 600 to 650 ms during emergence (Figure B) and again normalized to 449 ms after extubation. No other notable arrhythmias and no fluctuations in blood pressure, heart rate, or esCCO were observed. The patient's recovery and postoperative course was uneventful, and he was discharged the next day.

DISCUSSION

Although the cause of the QT prolongation in this patient was not clear, there are various types of congenital long QT syndrome, such as those with sensitivities to sympathetic or parasympathetic activity. Therefore, our general anesthetic goal in this patient was to quickly detect fluctuations in QTc along with heart rate and cardiac output.

Several anesthetic agents have the potential to prolong the QTc. Although barbiturates containing thiamylal can clinically prolong QTc,[5] thiopental reduces the transmural dispersion of repolarization (TDR). An increased TDR is associated with the onset of torsade de pointes.[6] On the other hand, no publications about the effects of thiamylal on TDR could be found using the PubMed database. Barbiturates such as thiopental could be beneficial in helping prevent torsade de pointes by reducing TDR, and

barbiturates have been used safely in patients with QT prolongation. Propofol does not modify TDR and can counteract QT prolongation, although its effects on QTc are conflicting.[6] However, thiamylal but not thiopental has been used in our hospital, and propofol availability was limited due to priority use in mechanically ventilated patients with COVID-19 when this patient was scheduled. Although volatile anesthetics can prolong the QTc, sevoflurane has less effect on QT prolongation in younger patients[7] and appears not to affect the TDR.[8] Opioid analgesics can suppress QT prolongation during intubation.[6] Rocuronium does not extend the QTc, and midazolam does not impact either the QTc or the TDR.[6] The epinephrine equivalent to 0.7 μg/kg administered during local anesthesia did not prolong the QTc in this patient, whereas its prolongation was reported after the injection of lidocaine containing epinephrine equivalent to 0.9 μg/kg in a child with QT prolongation.[9]

In this patient, it is possible that the QT prolongation noted during induction and intubation was primarily triggered by thiamylal and activation of the sympathetic nervous system. During anesthesia of children with LVNC and QT prolongation, they must be monitored for hemodynamic fluctuations, and careful consideration must be given to planning for the possible occurrence of arrhythmias as well as their respective management. The monitoring of intraoperative QTc and esCCO[10] might be useful for reducing the risk of arrhythmias and quickly detecting hemodynamic fluctuations.

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