Perioperative Management of a Patient With Cornelia de Lange Syndrome and Tetralogy of Fallot

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This is a case report of a 21-year-old male patient with Cornelia de Lange syndrome (CdL) and unrepaired tetralogy of Fallot scheduled for dental treatment under general anesthesia. Anticipated dental care consisted of restorative treatment and extractions. Surgical correction of the patient's congenital cardiac abnormalities had not occurred by the time of dental treatment. As such, the developed anesthetic plan included the following goals: prevention of any anoxic episodes or spell and preparation for difficult airway management due to micrognathia secondary to CdL. To help ensure adequacy of oxygenation during induction, the normal anesthetic face mask was specially modified with a hole to permit use of the fiberoptic scope during induction and intubation. With preoperative consultation involving a cardiologist as well as the use of the modified mask, general anesthesia was safely administered without any complications.

Key Words: Cornelia de Lange syndrome; Tetralogy of Fallot; General anesthesia; Fiberoptic intubation; Face mask modification.

Cornelia de Lange syndrome (CdL) is characterized by growth disturbances (short stature, micrognathia), intellectual disability, congenital heart disease (ventricular septal defect [VSD], atrial septal defect, pulmonary stenosis, tetralogy of Fallot [TOF]) and multiple other physical malformations (abnormalities of bones in the arms, hands, and fingers, low-set ears, small and upturned nose). Here is a report of a case involving general anesthesia for a 21-year-old patient with CdL and unrepaired TOF, further complicated by micrognathia. Main points of anesthesia management in this case were preventing anoxic episodes and managing an airway complicated by micrognathia.

CASE REPORT

A 21-year old male patient (height 131 cm; weight 23 kg; body mass index 13.4 kg/m²) was scheduled for dental

treatment under general anesthesia due to extensive dental caries and poor compliance secondary to intellectual disability. He was formally diagnosed with TOF following cardiac catheterization at 1 year of age, which is a combination of 4 congenital cardiac malformations: VSD, overriding aorta, pulmonary valve stenosis, and right ventricular hypertrophy. When providing anesthesia for patients with TOF, attention should be paid to prevent anoxic episodes or "tet spells," maintain ideal hemodynamics to prevent shunting, and prevent infectious endocarditis. Additionally, the patient reported taking carteolol hydrochloride to maintain his stable cardiac status.

A cardiologist was consulted regarding assessment of the patient's cardiac condition prior to the operation. ECG showed right ventricular hypertrophy, and echocardiography revealed right ventricular flow tract obstruction, VSD (maximum diameter of 15 mm), bidirectional shunt, and a left ventricular ejection fraction of 62%. Pulmonary valve diameter was 11.3 mm, and the right ventricular to pulmonary arterial pressure gradient was 64 mm Hg. The stated severity of the patient's TOF was deemed minor, and the patient reported no history of any anoxic spells in the past.

The patient was unable to follow instructions at the time of the physical examination due to his intellectual disability. As such, it was not feasible to adequately

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Figure 1. Anesthesia face mask modified with a hole.

assess key physical findings such as neck extension and his range of mouth opening due to the limited extent of the preoperative airway examination.

The main points of anesthesia management in this case were preventing anoxic episodes and managing an airway complicated by micrognathia. Use of a fiberoptic scope was anticipated secondary to the patient's micrognathia. The anesthetic plan included a slow mask induction using sevoflurane, along with nitrous oxide and oxygen, followed by gentle intubation assisted with a fiberoptic scope. Beforehand a standard anesthesia face mask was prepared with a hole (Figure 1), which permitted continued administration of the inhaled anesthetic gases, ensuring adequate anesthetic depth even during operating fiberoptic intubation.

After placement of standard anesthetic monitors (NIBP, SpO₂, ECG, EtCO₂), general anesthesia was induced with nitrous oxide (4 l/min), oxygen (2 l/min), and sevoflurane (2%). After unconsciousness was achieved, an intravenous line was established in the patient's upper extremity. The hole in the anesthesia face mask was temporarily covered with tape at the start of inhalation induction to prevent anesthetic gases from leaking out. After adequate depth of anesthesia was ensured, his limited mouth opening due to the micrognathia was confirmed at 20 mm, and the tape was removed from the hole. A 6.0 mm preformed nasal endotracheal tube was passed through the hole of the mask and into the left nostril. The nasal tube was passed easily through the nasal cavity. Fiberoptic intubation could then be successfully performed while the modified mask was connected to the anesthesia circuit, enabling continuous delivery of sevoflurane, nitrous oxide, and oxygen to the patient during use of the fiberoptic scope and subsequent intubation. It must be appreciated that the modified mask could not provide positive pressure

Figure 2. The mask during intubation with the fiberoptic scope. While maintaining spontaneous breathing, the endotracheal tube with the connector removed beforehand was passed through the hole in the mask and into the nostril. The fiberoptic scope was then inserted through the endotracheal tube. After successful intubation, endotracheal tube was reconnected to the anesthetic circuit. This maneuver allowed administration of anesthetic gases throughout the fiberoptic intubation.

ventilation due to the predrilled hole. The patient slightly moved and coughed when nasotracheal tube was passed successfully through the vocal cords. Successful intubation was confirmed with appropriate continued detection of end-tidal CO₂.

General anesthesia was maintained throughout the rest of the case using propofol (5.5–6.5 mg/kg/h; 92–108 µg/kg/min), remifentanyl (0.1–0.25 µg/kg/min), air (5 l/min), and oxygen (1 l/min). The total volume of Ringer's acetate solution with 5% dextrose infused was 500 mL. General anesthesia was well maintained throughout the duration of the case with blood pressure, heart rate, and arterial oxygen saturation remaining stable. The patient was extubated after emergence while spontaneously breathing without difficulty, and no perioperative complications were noted.

DISCUSSION

For patients with TOF, anoxic episodes or tet spells may be caused by stress, which can lead to an increase in right-to-left shunting of blood flow secondary to pulmonary spasm.¹ Rough laryngoscopy could cause pharyngeal or laryngeal injury with resulting edema of the glottic anatomy. Further complicating this patient's anesthetic management was micrognathia. Therefore, a slow mask induction while maintaining spontaneous ventilation was utilized, along with a gentle fiberoptic

intubation. Care was taken to avoid intubation at lighter levels of anesthesia because stimulation of the airway can trigger an anoxic spell. Use of the modified face mask with the hole allowed delivery of the anesthetic gases throughout induction, even during use of the fiberoptic scope. Using this device, nasal intubation was successfully established (Figure 2) with minimal stimulation of the patient's airway. Sevoflurane concentration could be easily controlled, maintaining its concentration at 1% to 2% without obstructing spontaneous breathing during manipulation of the fiberoptic scope. Propofol, used as the primary intravenous maintenance agent, further contributed to reducing the incidence of emergence agitation.²

It is critical to develop individualized anesthetic plans that address the potential difficulties that may arise during the provision of anesthesia. These plans must adequately address the patient's comorbidities. In this case, modification of the traditional anesthesia face mask permitted uninterrupted delivery of inhaled anesthetic gases during fiberoptic intubation. Addition-

ally, it may be appropriate to consider various other steps such as consultation with medical specialists (cardiologists) during the preoperative assessment phase of providing anesthetic care for patients when indicated.

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