The Anesthetic Management for a Patient With Trisomy 13

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Trisomy 13 is a chromosomal disorder that occurs in complete or partial mosaic forms. It is characterized by central apnea, mental retardation, seizure and congenital heart disease. The survival of the patients with trisomy 13 is the majority dying before one month. Trisomy 13 is the worst life prognosis among all trisomy syndromes. It is reported the cause of death is central apnea. Special needs patients with mental retardation are recognized to have poorer oral health condition. Oral health related quality of life reflects daily activity and well-being. Dental treatment under general anesthesia is sometimes an option for such patients. This patient had received ventricular septal defect closure surgery at 2-year-old. In addition, he had mental retardation and seizure. Dental treatment had been completed without any cerebral and cardiovascular events under non-invasive monitoring with not only cardiac electric velocimetry, but also epileptogenic activity. In addition, postoperative respiratory condition was maintained stable in room air.

Key Words: Trisomy 13; General anesthesia; Non-invasive cardiac function monitor; Congenital heart disease.

Patients with special needs often present with intellectual disability such as reduced cognitive skills and impaired cooperation with resultant decreased quality of life.[1] Dental caries is a common condition in these patients. Pain due to dental caries may cause eating problems that may not only affect daily activity but may lead to life-threating complications of untreated oral infection. However, it is difficult for the patients to receive dental treatment in the usual office setting. General anesthesia is frequently requested for their dental treatment.

Patients with trisomy 13 often have central sleep apnea, intellectual disability, seizure, cleft palate, microcephaly, and congenital heart disease such as patent ductus arteriosus (PDA) and ventricular septal defect (VSD).[2,3] The anesthetic management of these patients has only been sporadically reported.[3] We report the anesthetic management for an 18-year-old male with trisomy 13 who required dental treatment for caries.

CASE PRESENTATION

The patient was an 18-year-old male, 160 cm (63 in) in height and 41 kg in weight. He was diagnosed with trisomy 13 at birth. Multiple caries were diagnosed at dental examination when the mother observed his difficulty eating. Due to his inability to cooperate, dental treatment was scheduled under general anesthesia. He had received ventricular septal defect closure surgery at 2 years of age. Preoperative echocardiography was unremarkable. Ejection fraction was 65%. A 12-lead electrocardiogram showed normal sinus rhythm. Chest radiograph was within normal limit. On admission, his heart rate (HR) was 95 beats per minute (bpm), and blood pressure (BP) was 110/70 mmHg, SpO_2 was 100% on room air. Auscultation of heart and lungs was within normal limits. His neurological status was poor with severe mental retardation. He could articulate a few words, and we could not communicate with him. The clinical assessment of his airway was difficult. He could take a sitting position, but would not accept dental examination or treatment. He was taking daily antiepileptic drugs (sodium valproate) and had no seizures for more than 2 years. In addition, there was no history of apnea that was apparent.

On the day of dental treatment, no premedication was given and he was transferred to the operating room. After pulse oximeter placement (100%), anesthesia was induced with inhalation of sevoflurane 5% in nitrous oxide 30% and oxygen. After loss of consciousness, mask ventilation was easy. Electrocardiogram (sinus rhythm), BP (105/75 mmHg), HR (87 bpm) and Bispectral Index (BIS value: 8–20, spectral edge frequency: 10–20 Hz, signal quality index: 95%) monitoring were started. In addition, cardiac status was monitored using Aesculon®. Atropine 0.25 mg, fentanyl 100 μg, and rocuronium 25 mg were adminis-

Received May 21, 2016; accepted for publication September 8, 2016.

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Anesth Prog 64:162–164 2017 | DOI 10.2344/anpr-64-02-09

tered after peripheral intravenous access was obtained. Intubation was easily carried out with a 6.5-mm nasotracheal tube. Anesthesia was maintained with isoflurane 1–1.5%, in air and oxygen. During anesthesia, BP was maintained at 80–110/50–70 mmHg, HR was 75–95 bpm, SpO_2 was 98–100%, and $EtCO_2$ was 35–40 mmHg. Hemodynamic parameters via Aesculon® were cardiac output 2.9–3.7 L/min, stroke volume variation 9–11%, and stroke volume 35–48 mL. BIS value was maintained between 40–58 with spectral edge frequency 10–15 Hz and signal quality index 95%. A total of 1.8 mL of 2% lidocaine containing 1 : 200,000 epinephrine was injected for dental treatment. Tooth extraction and restorative treatment for caries was completed uneventfully. The operation was completed in 350 minutes without any surgical and/or anesthetic problems. There was minimal blood loss during operation, and he received a total of 1050 mL acetated Ringer's solution with 1% glucose. In addition, total urine volume was 400 mL. He was extubated fully awake and did well postoperatively with stable vital signs. Following extubation, his hemodynamic condition and oxygen saturation was stable (BP: 110/58 mmHg, HR: 101 bpm, SpO_2: 98%). Postoperative SpO_2 98–100% was maintained during the postoperative period in room air. The patient was admitted for overnight observation and monitoring in our dental hospital.

DISCUSSION

Patients with special needs tend to have complex dental problems and poor oral health status.[4] This is especially true for those with severe intellectual disability who may often have difficulty eating due to toothache. Left untreated, these conditions may develop severe, even life-threating infections. Dental treatment under general anesthesia can be a great support for these patients.

Trisomy 13 is the third most viable trisomy in autosomes, following trisomy 21 and trisomy 18.[5,6] It is estimated to occur in 1 : 5000 to 20,000 live births with many conceptions not surviving to birth.[2,5] Death can occur early in life. The clinical features are characterized by central apnea, cardiac defects, seizure disorder, and severe mental retardation.[2,5,6] One of the major causes of death is due to central apnea.

Anesthetic management includes seizure prevention, maintenance of stable hemodynamics, particularly in those with ongoing cardiac compromise, and close postoperative respiratory monitoring for apnea. Sevoflurane is a widely used volatile anesthetic, especially for inhalation induction. However, it may induce the epileptogenic activity, particularly with hypocarbia.[7] Isoflurane was used for maintenance of anesthesia to decrease possible seizure occurrence, which is common in patients with trisomy 13. In this case, we monitored epileptogenic activity by using BIS as has been reported.[2,3] As hyperventilation may decrease cerebral blood flow leading to seizure, we maintained $EtCO_2$ within normal levels. No seizure activity was observed via the BIS or postoperatively.

Congenital heart disease is frequently associated with trisomy 13, but usually this is noncyanotic with VSD and PDA most common.[2,3] In this case, we used the Aesculon® to follow trends in hemodynamic parameters, although preoperative assessment including echocardiography, electrocardiogram, and chest radiograph were within normal limit.[8,9] This device is a noninvasive cardiac function monitor using electrical velocimetry method based on the premise that the orientation of the erythrocytes in the aorta changes quickly from random alignment to alignment in the direction of blood flow upon opening of the aortic valve.

Concerning central sleep apnea, it may exist alone or in combination with obstructive sleep apnea syndrome and results in abnormal breathing during sleep. Primary central sleep apnea is a rare condition, and the etiology is not entirely understood. In this case, we were concerned that central apnea might be triggered by prolonged anesthetic action. We avoided administering long-acting anesthetics, and postoperative SpO_2 was monitored until the next morning.

CONCLUSION

Dental caries has many consequences including inability to eat, pain, and potentially life-threating infection. Patients with special needs, especially those with intellectual disability, frequently require general anesthesia for dental and oral surgery. We present a case of general anesthesia of a patient with trisomy 13, in which we paid special attention to seizure avoidance, stable hemodynamics, and monitoring of the postoperative respiratory condition.

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