Egyptian Journal of Anaesthesia (2017) 33, 213–215

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Egyptian Society of Anesthesiologists

Egyptian Journal of Anaesthesia

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Case report

The anesthetic management for a special needs patient with trisomy 18 accompanying untreated tetralogy of Fallot

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Received 30 April 2016; accepted 15 September 2016
Available online 30 September 2016

KEYWORDS

Trisomy 18;
Untreated TOF;
General anesthesia

Abstract 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 often an option for such patients.

Trisomy 18 is characterized by congenital heart disease, craniofacial abnormality and mental retardation. Congenital heart disease can be greater risk during anesthesia. In the case of trisomy 18 with untreated tetralogy of Fallot, especially right-to-left shunting and/or pulmonary artery stenosis may reduce pulmonary blood flow, and may develop life-threatening hypoxemia.

We anesthetized a patient with trisomy 18 accompanying untreated tetralogy of Fallot for dental treatment. The hemodynamics including cardiac output has been monitored non-invasively using electrical velocimetry method. Its systemic vascular resistance and pulmonary vascular resistance were maintained appropriately, and dental treatments were successfully completed.

1. Introduction

Some special needs patients have intellectual disability including deficient cognitive skills and impaired cooperation, and their quality of life might be decreased [1]. Dental caries is a common oral problem in these patients. Pain due to dental caries may easily cause eating disorder, which may not only affect their daily activity but also be sometimes life-threatening. However, it is difficult for the patients to receive dental treatment in usual settings. General anesthesia is often requested for their dental treatment [2].

Patients with trisomy 18 often have mental retardation and congenital heart disease [3]. In these patients, dental caries may easily develop systemic infection and might be mortal. General anesthesia is usually required for their dental treatment. However, the mortality rate is high around the perioperative period

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Peer review under responsibility of Egyptian Society of Anesthesiologists.

http://dx.doi.org/10.1016/j.egja.2016.09.001

in trisomy 18 patients. On the other hand, very few reports have examined the anesthetic management for long survival patients [4–6]. Therefore, untreated TOF can be greater risk for general anesthesia. We experienced an anesthetic management for a 12-year-old girl with trisomy 18 accompanying untreated TOF.

2. Case report

The patient was a 12-year-old girl, 112 cm in height and 16 kg in weight. She was diagnosed as trisomy 18 at birth. Simultaneously she had tetralogy of Fallot (TOF), but it had been remained untreated. Her mother found her multiple dental caries as her eating disorder had been observed. Dental treatment was scheduled for dental caries under general anesthesia.

Preoperative echocardiography revealed TOF with aorta overriding, a small ventricular septal defect with bidirectional shunting, and pulmonary stenosis (PS) (pressure gradient 39 mmHg). The right ventricle was moderately dilated and hypertrophied. Ejection fraction was 74%. A 12-lead electrocardiogram was sinus rhythm, and showed right axis deviation and incomplete right bundle branch block. Chest X-ray was within normal limits. In laboratory investigations Hb concentration was 19.4 g/dL, and Hct was 56.7%. On admission, her heart rate (HR) was 95 bpm, and blood pressure (BP) was 110/70 mmHg. SpO_2 was 80% in room air, but it often decreased to around 70% when she was agitated. A loud ejection systolic murmur was noted in the pulmonary area. She regularly received follow-up observation. Her neurological status was poor with severe mental retardation. She could talk only a few words, but we could not communicate with her. She could take sitting position, but could not stand by herself. She was given daily antiepileptic drugs, and had no attack of seizure for more than two years. The clinical assessment of her airway was difficult.

On the day of dental treatment, no premedication was given, and she was transferred to the operating room with a 24-gage peripheral venous access at the dorsum of the left hand. After the application of standard non-invasive monitoring, including ECG (sinus rhythm), SpO_2 (78%), HR (96 bpm) and NIBP (100/60 mmHg), anesthesia was induced with intravenous propofol 2–5 mg/kg/hr, fentanyl 100 μg titrated to hemodynamics in 100% oxygen. After the loss of consciousness, mask ventilation was not difficult. Rocuronium 10 mg was also given intravenously to facilitate tracheal intubation. We could smoothly intubate by using laryngoscope with a 4.5-mm nasotracheal tube. Anesthesia was maintained with propofol and intermittent bolus of fentanyl. FiO_2 was maintained 0.7–1.0. A 24-gage catheter was inserted into the left radial artery for BP monitoring. In addition, cardiac status was monitored by using Aesculon®. During anesthesia BP was maintained at 80–110/50–70 mmHg, and SpO_2 was 70–80%. HR was 95–140 bpm, cardiac output (CO) was 1.9–3.2 L/min, stroke volume (SVV) was 9–11% and stroke volume (SV) was 15–25 ml. Intraoperative saturation ranged from 90% to 95%. Total 5.4 ml of 2% lidocaine containing 1:200.000 adrenaline was injected for dental treatment. Two episodes of hypotension (systolic BP $<$ 80 mmHg) during dental treatment were treated by phenylephrine 10 μg. Intra-oral X-ray and the dental treatment for teeth decay were completed uneventfully. The operation was completed in 67 min without any surgical and/or anesthetic problems. There was minimal blood loss during operation and she received a total of 500 mL lactated Ringer's solution. She emerged from general anesthesia in 10 min with after stopping inhalation of sevoflurane. She was extubated after confirming sufficient spontaneous respiration. After extubation her respiratory and hemodynamic conditions were stable (NIBP: 110/58 mmHg, HR: 101/min, SpO_2: 80%).

3. Discussion

Special needs patients tend to have complex dental problems and poor oral health status, since it is difficult for them to receive preventive dental care and to understand its importance. In particular, special needs patients with severe mental problems may often stop eating because of toothache, and it may develop to life-threatening condition. Dental treatment under general anesthesia could be a great support for those patients. On the other hand, we faced on the dilemma that those patients often have severe cardiovascular and/or pulmonary problems.

Trisomy 18 was first described by Edwards in the 1960s [4–6]. The clinical features of trisomy 18 were characterized by cardiac defects, convulsion, and growth deficiency such as micrognathia, facial clefts, and mental retardation [4–6]. It is estimated 1/8000, but most of them die in fetal life [5,6]. Approximately 50% of babies with trisomy 18 could live longer than one week, and only five to 10% of babies survive beyond the first year [6]. The sex ratio is usually four girls for every boy [4]. One of the major causes of death is congenital heart defect. Survival without surgical treatment for cardiac defect is exceedingly rare but reported [5,6]. It might report that her unusual longevity is owed to a well-balanced intracardiac shunt and good pulmonary blood flow via large branch pulmonary arteries with enough restriction to avoid overcirculation [6].

In general anesthesia with severe heart disease patients, it is important to monitor strict hemodynamic change, i.e. central venous pressure and/or pulmonary artery wedge pressure. However, insertion of central venous catheter or pulmonary artery catheter is by far invasive procedure comparing to dental treatment. In this case, cardiac function was monitored by Aesculon® using electrical velocimetry monitor, which is non-invasive cardiac function monitor [7,8]. This monitor is based on the premise that the orientation of the erythrocytes in the aorta changes quickly from random to alignment in the direction of blood flow upon opening of the aortic valve.

The goals of anesthetic management for the patient with untreated TOF were to maintain in systemic vascular resistance (SVR) and to avoid decrease in pulmonary vascular resistance (PVR) [3.5]. It is important to maintain SVR, since SVR was relative to PVR minimizes right-to-left shunting [5]. In this case, during anesthesia, transfusion was administered with reference to urine volume and hemodynamics changes such as CO, SV and SVV. Intermittent phenylephrine boluses were administered to avoid to decrease less than 80 mmHg. In addition, FiO_2 was maintained with 0.7–1.0 due to increasing pulmonary blood flow [4,5]. Since the crying children with TOF might easily fall in the anoxic spell, before induction of anesthesia intravenous access was obtained, and propofol and fentanyl were administered as a titration to maintain

stable hemodynamics during dental treatment. In addition, no further stimulation is allowed until she spontaneously wake up.

4. Conclusion

Tooth pain due to dental caries occurs life-threatening. It is necessary for special needs patients with eating disorder to receive dental treatment under general anesthesia for better life. In general anesthesia, it is useful for severe cardiac disease patient to use noninvasive monitoring.

Conflict of interest

None declared.

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