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Management of difficult airway in pediatric patients with right ventricular outflow tract obstruction

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Abstract
We present two cases of difficult airway management for patients with Pierre Robin syndrome and right ventricular outflow tract obstruction in infants. To prevent the exacerbation of right ventricular outflow tract obstruction, adequate oxygenation and ventilation are mandatory in this population. This rule needs to be followed even while dealing with a difficult airway. Depending on the prediction of mask ventilation capability, we took two different approaches to difficult airway. In the first case, we fiberoptically intubated the patient while allowing him to breathe spontaneously with the aid of a nasopharyngeal airway under deep sedation. In the second case, we fiberoptically intubated the patient through a laryngeal mask airway while controlling ventilation. Through both cases, we highlight options of difficult airway management in the pediatric population. Although we can approach a difficult airway with or without spontaneous breathing, the important point is how we will prepare the methods to oxygenate and ventilate patients throughout the procedure. Patients with difficult airway and right ventricular outflow tract obstruction are good examples to make us realize this point.

Key words Pierre Robin syndrome · Difficult airway · Right ventricular outflow tract obstruction · Cyanotic episode

Introduction

Pierre Robin syndrome is a well-known condition associated with micrognathia, glossoptosis, and cleft palate [1]. It often challenges medical practitioners with difficult mask ventilation or difficult intubation. Patients with Pierre Robin syndrome often have congenital heart disease. According to Pearl [2], congenital heart diseases were observed in about 20% of this population, and ventricular septal defect (VSD), patent ductus arteriosus, and atrial septal defect were the most common cardiac lesions among them. The prevalence of right ventricular outflow tract obstruction (RVOTO), as illustrated in the patients we report here, is not clear, but Pearl [2] reported that about 10% of patients with Pierre Robin syndrome and congenital heart disease had tetralogy of Fallot (TOF).

Hypoxia and hypercarbia pose serious problems to patients with RVOTO by further increasing pulmonary vascular resistance (PVR). Patients with intracardiac shunt demonstrate quicker desaturation from the increase of right-to-left shunt. Patients with no intracardiac shunt will develop pressure overload of the right heart, resulting in low cardiac output from right heart failure. Therefore, a particularly well-prepared anesthetic plan is necessary to manage patients with a difficult airway with RVOTO. Additionally, pediatric patients usually have more limited reserve than adults because of (1) their increased oxygen demand and (2) lower functional residual capacity, because they lose their expiratory braking following the induction of general anesthesia [3]. As a result, decompensation can occur faster than it does in adults.

Here we present two cases of successful fiberoptic intubation for patients with Pierre Robin syndrome and RVOTO. Based on the prediction of mask ventilation feasibility, we used two different strategies, as described below. These may suggest one way of airway approach for this patient population.

Case reports

Case 1

A 2-month-old, 3.0-kg baby boy, formally born at 34 weeks, was diagnosed with Pierre Robin syndrome, double-chamber right ventricle (DCRV), and a large

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membranous VSD. He had had frequent episodes of cyanosis since birth. Dynamic RVOT obstruction with increased right-to-left shunt across the VSD was considered to be a major cause of the cyanosis, rather than his airway obstruction.

He was transferred to our institution for further management. His hemodynamic profile was stable and peripheral oxygen saturation ($S_{P_{O_2}}$) was usually in the high 90s%, but it dropped to the 60s% with agitation. Cyanotic episodes were successfully treated with intravenous morphine. Transthoracic echocardiogram confirmed a moderate-sized VSD, a severe DCRV with a maximum instantaneous pressure gradient of 95 mmHg, and patent foramen ovalis (PFO). He presented to the operating room for DCRV repair. Vital signs included blood pressure (BP), 80/40 mmHg; heart rate (HR), 120 beats·min^{-1}; $S_{P_{O_2}}$, 96%. Incremental doses of midazolam (total 0.2 mg) and morphine (total 0.75 mg) were given intravenously until he was deeply sedated. Both of his nostrils were prepared with oxymetazoline spray (topical alpha-agonist decongestant). Scopolamine 0.03 mg was administered intravenously for drying out the mouth. Because substernal retraction was noted with breathing following deep sedation, a 3.0-mm uncuffed endotracheal tube (ETT) was inserted from one nostril as a nasopharyngeal airway, which was connected to the breathing circuit. With the insertion of the nasopharyngeal airway, substernal retraction was no longer noted. We prepared a laryngeal mask airway (LMA) #1 as a backup in case the airway became obstructed and ventilation could not be managed manually or by using the nasal pharyngeal airway. Otorhinolaryngology (ORL) surgeons were on stand-by in the room. We performed fiberoptic intubation (outer diameter 2.2 mm) from the other nostril. After visualization of the vocal cords, 1% lidocaine was injected through the scope. The trachea was intubated with a 3.0-mm uncuffed ETT without any problems. Spontaneous breathing was maintained throughout this course. Throughout this process, vital signs were stable. Following intubation, he was paralyzed with vecuronium. Anesthesia was maintained with 0.25%–0.5% isoflurane and fentanyl (total 250 μg). Surgery included patch closure of the VSD via infundibulotomy and resection of the right ventricle muscle bundle. The PFO was left open. Epicardial echocardiogram revealed a small residual VSD with a pressure gradient of 10–15 mmHg across the RVOT with 1/2–2/3 systemic RV pressure. Postoperatively he was transported to the intensive care unit (ICU) intubated. On postoperative day 5, he was extubated. Cyanotic episodes were no longer noted. He was transferred back to his original hospital.

Case 2

A 2-month-old boy, weighing 3.5 kg, with Pierre Robin syndrome, status postsurgical repair of coarctation of aorta, and pulmonary stenosis, was found to have moderate to severe re-coarctation of aorta and moderate pulmonary valve stenosis on a transthoracic echocardiogram at office visit. He was scheduled to have a balloon dilation of coarctation and pulmonary valve by cardiac catheterization. In the previous surgery, he had been intubated by the ORL surgeon following airway examination.

Because there had been no problem with mask airway ventilation in the previous surgery, he was induced with etomidate 1 mg and mask-ventilated. After paralysis with cis-atracurium 0.5 mg, direct laryngoscopy was attempted, without success. We placed a disposable LMA #1 without any problem and maintained adequate oxygenation and ventilation. Then we intubated fiberoptically (outer diameter 2.2 mm) using a long uncuffed 3.0-mm ETT through the LMA via the adaptor connected to the LMA. To facilitate the passage of the ETT through the LMA, we cut off the slits at the LMA opening. Assisted ventilation was continued throughout this procedure. The LMA was left in place. The anesthesia was maintained with 0.5%–1.2% isoflurane and fentanyl (total 10 μg). The procedure was uneventful. He was extubated at the end of the procedure and transferred back to the regular ward.

Discussion

Difficulties with intubation and ventilation contribute significantly to morbidity and mortality with anesthesia. In fact, difficult intubation is the second most frequent primary damaging event leading to malpractice claims [4]. The American Society of Anesthesiologists (ASA) developed a practice algorithm for difficult airway management to assist practitioners in making decisions [5]. However, its focus is completely on the airway, and the ASA algorithm does not address the importance of disease-specific ways of management. Furthermore, providing oxygen is greatly stressed, while dealing with a difficult airway, but with adequate ventilation to keep normocapnia is not.

Here we presented two cases of difficult airway management for patients with RVOTO. In general, both hypoxia and hypercarbia increase PVR. These will further worsen the degree of RVOTO. Because exacerbation of RVOTO can impair oxygen saturation or right ventricular function, methods to provide adequate ventilation, as well as oxygenation, are especially important while dealing with difficult airway in this population.

We took two different approaches to deal with presumed or known difficult airway, based on the knowledge of the patients' mask ventilation capability. In both patients, the presumed or known difficult airways were from Pierre Robin syndrome. It is a well-known condition that often challenges us with difficult mask ventilation or difficult intubation. The first patient had DCRV. Primary DCRV is a rare congenital anomaly consisting of one or more anomalous muscle bundles that divide the right ventricle into a proximal high-pressure chamber and a distal low-pressure chamber [6,7]. The natural history of this lesion is not well defined, but there is a high association with other cardiac lesions, especially membranous-type VSD, and a tendency for the obstruction to progress over time. This patient's physiology was very similar to that of TOF. Adequate hydration and keeping patients calm are very important to prevent cyanotic episodes. Because manipulating the airway when the patient is awake or under light sedation could potentially cause agitation and a cyanotic episode, we chose to titrate sedation slowly to make the patient sedated deeply enough to allow airway manipulation. We also paid special attention to maintaining spontaneous breathing, because we were not sure about the feasibility of mask ventilation. A nasopharyngeal airway was used to lessen airway obstruction following deep sedation. We also tried to avoid iatrogenic tachycardia because it would have further worsened RVOTO. Therefore we chose scopolamine as an antisialogogue because of its minimal effect on heart rate. In the second case, we induced and paralyzed the patient because we knew that he had been mask-ventilated easily in the past. We placed an LMA after a failed direct laryngoscopy and intubated the trachea through the LMA fiberoptically, while maintaining ventilation [8]. Additionally, if we had tried and had been able to mask-ventilate the patient in the first case, we could have induced him, and placed an LMA through which the trachea would have been intubated, as was done in the second patient.

In conclusion, the two cases presented here illustrate one approach to difficult airways for pediatric patients with congenital heart disease. Although we could approach a difficult airway with or without spontaneous breathing, as we showed here, the important point here is how we should prepare the methods to oxygenate and ventilate patients throughout the procedure. Patients with difficult airway and RVOTO are good examples to make us realize this point.

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