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

KOICHI YUKI and ALFONSO CASTA

Department of Anesthesiology, Perioperative and Pain Medicine, Children’s Hospital Boston and Harvard Medical School, Boston, MA, USA

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 disease 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
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 ($\text{SpO}_2$) 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$^{-1}$; $\text{SpO}_2$, 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.