Anesthetic management of double chamber right ventricle repair for a patient with Pierre Robin syndrome

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Introduction: Difficult airway with congenital heart disease poses a difficult situation to anesthesiologists. We will present the case of double chamber right ventricle repair for a patient with Pierre Robin syndrome.

Case report: 2 month old, 3.0kg babyboy (former 34 weeker) with Pierre-Robin sequence, double chamber right ventricle (DORV) , large membraneous VSD, was initially managed at outside hospital because of his frequent cyanosis. Because his increased episodes of cyanosis and upper airway obstruction, tongue-lip adhesion was done at outside hospital. Anesthesia was induced with 10mg propofol, LMA#1 was placed without any difficulty. Mask ventilation feasibility was not documented. Intraoperative course was uneventful and LMA was removed at the end per record. Despite the surgery, episodes of cyanosis continued, which was always associated with agitation. Right ventricular outflow tract (RVOT) dynamic obstruction with left to right shunt across VSD was considered to be a major cause of cyanosis.
He was transferred to our institution for further management. Preoperative echocardiogram showed moderate sized VSD, severe DORV with maximum instantaneous gradient of 95mmHg, PFO. He presented to operating room with one peripheral IV. The decision was made to do sedated fiberoptic intubation, maintaining spontaneous breathing. IV midazolam and morphine was given in increment doses until deeply sedated. Bilateral nostril was prepped with afrin spray. IV scopolamine 0.01mg/kg was given to dry out the mouth. One 3.0 uncuff ETT was inserted from one nostril as nasopharyngeal airway, which was connected to breathing circuit and from another nostril, fiberoptic intubation was performed relatively easily. 3.0 uncuffed ETT was inserted. Then he was paralyzed with vecuronium. Left radial arterial line, right internal jugular central line was inserted for monitoring. Surgery included patch closure of VSD via infundibulotomy and resection of right ventricle muscle bundle. PFO was left open. He was stable intraoperatively. Postoperative epicardial echocardiogram revealed small residual VSD, 10-15mmHg gradient across RVOT. ½-2/3 systemic RV pressure. He came off CPB with any problems. POD#5 he was extubated. He was transferred back to his original hospital.

Discussion: we presented a case of possible difficult airway with congenital heart disease. Incidence of difficult airway with congenital heart disease is unknown, and this population gives us unique problems. For infant with possible difficult airway, typically awake or slightly sedated fiberoptic or laryngoscopy will be given a consideration. This patient had severe RVOT obstruction with muscle bundles, and had multiple episodes of “cyanotic spell” like tetralogy of Fallot. To avoid cyanotic episode, good hydration and keeping patient calm is very important. So manipulating airway when the patient was awake or in light sedation was not optimal for this patient. For this reason,we chosed to titrate sedation slowly to make patient deep enough to allow airway manipulation. Also we paid special attention to maintain spontaneous breathing, since we were not sure about the capability of mask ventilation. We prepared LMA as back up. ORL surgeons were stand-by in the room. We used scopolamine as antisialagogue because of its minimal effect on HR. Tachycardia can be detrimental with his severe RVOT obstruction. We placed one ETT tube from one nostril as nasopahyngeal airway to relieve airway obstruction and as a source of oxygen supply, since we wanted to avoid hypoventilation and resultant elevation of pulmonary vascular resistance, which can worsen R to L shunt. We performed fiberoptic intubation from

other nostril. After visualization of vocal cord, 1% lidocaine was injected through the scope around the vocal cord. The patient was fiberoptically intubated without problems. For potential spells, we prepared 5% albumin as volume expander, esmolol, and phenirephrine in the room. The rest of the procedure went smoothly. This case illustrates one way of approaching difficult airway with congenital heart disease. Also we need to put a great emphasis on multidisciplinary approach.