Difficult Intubation in a Child With Pierre Robin Sequence Associated With Stickler Syndrome Using a Glidescope and Gum Elastic Bougie

Mukesh Wadhwa, DO, Shiraz Yazdani, MD, Ashraf Farag, MD, Dennis Ho, DO
Texas Tech University Health Sciences Center, Lubbock, TX

Introduction

Pierre Robin sequence is a craniofacial condition. It is referred to as a sequence because it results as a series of events during a baby’s development in the womb. It is characterized by micrognathia, receding chin, cleft palate, and glossoptosis, making ventilation and intubation challenging. Children with this sequence may have feeding and respiratory difficulties. Approximately 40 percent of patients with Pierre Robin sequence also have Stickler syndrome. Stickler syndrome is a group of genetic disorders affecting connective tissue, specifically collagen. Stickler syndrome is characterized by distinctive facial abnormalities, ocular problems, hearing loss, and joint problems. The syndrome is thought to arise from a mutation of several collagen genes during fetal development. A characteristic feature of Stickler syndrome is a somewhat flattened facial appearance; this is caused by underdeveloped bones in the middle of the face, including the cheekbones and the bridge of the nose.

Case Report

A 15 month old male presented for cleft palate repair. He had a past medical history of Pierre Robin sequence associated with Stickler syndrome, asthma, GERD, and resolved ASD. He had a Nissen fundoplication and g-tube at birth and mandibular distraction at four months. On physical examination, he had micrognathia, posteriorly displaced tongue, cleft palate, normal mouth opening, and a Malampatti IV airway. Inhalation induction was performed in the OR. Direct laryngoscopy resulted in no visualization of vocal cords. Following easy ventilation, a Glidescope was inserted, resulting in a view of the posterior commissure. Due to inability to pass the endotracheal tube with just a view of the posterior commissure, a gum elastic bougie was utilized to assist in passing a 4.0 cuffed endotracheal tube. Intubation was determined successful upon confirmation by capnography and bilateral breath sounds. An attempt using fiberoptic intubation alone with inhalation induction was attempted but was unsuccessful due to difficulty with visualization secondary to difficult airway anatomy.

Discussion

Children with congenital abnormalities warrant special consideration when approaching the topic of securing the airway. They often present for surgical procedures of the oropharynx such as our patient, who presented for cleft palate repair. Their craniofacial abnormalities lead to many unique and potentially challenging airway scenarios. A thorough history and physical examination is essential, including review of previous anesthetic records to ascertain what may have been previously successful.

Several techniques have been demonstrated in view of the difficulty in intubation of patients with Pierre Robin sequence. One involved the use of the paraglottal approach to laryngoscopy in conjunction with use of a gum elastic bougie, when conventional laryngoscopy failed to obtain a grade I or II laryngoscopic view. Another technique involved retrograde nasal intubation. Lastly, a technique involved a combination of Glidescope and fiberoptic bronchoscope. The use of combination of a Glidescope with a fiberoptic bronchoscope or gum elastic bougie allows the ease of placement of an endotracheal tube along the curvature of the Glidescope. The advantage of the Glidescope is superior glottic views, although the sharp curvature of the blade may make passing an endotracheal tube difficult. This difficulty is attenuated by using a gum elastic bougie. The endotracheal tube can then easily be passed over the gum elastic bougie and viewed as it enters the trachea. Care must be taken to avoid tracheal trauma with the rigid gum elastic bougie.

References


The Pierre Robin sequence includes a U- or V-shaped cleft palate with relative macroglossia due to the smaller lower jaw. This image is an example of such an airway. The structure seen in the back of the oropharynx is a nasogastric tube.

Image of a child displaying typical features of Stickler Syndrome with Pierre Robin sequence.

One of the characteristic features is a somewhat flattened facial appearance caused by underdeveloped bones in the middle face, including the cheekbones and the bridge of the nose.