Anesthesia for an Infant with Pierre Robin Sequence and Pyloric Stenosis

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Goals:
1. Consider the conflicting anesthetic goals associated with anesthetizing infants with Pierre Robin Sequence and pyloric stenosis.
2. Formulate and justify an anesthetic plan to safely anesthetize the uncooperative infant with a full stomach and a difficult airway.
3. Discuss the potential risks and benefits of alternative techniques.

Case:
A six week old, full term male infant presents to the hospital with a history of several days of projectile, non-bilious vomiting. The child was diagnosed with Pierre Robin sequence shortly after birth based on severe micrognathia and associated large, midline cleft palate. He has required prone sleeping and feeding to maintain his airway but no invasive airway management. He has also a diagnosis of gastro-esophageal reflux but is otherwise healthy. Based on his presenting symptoms, abdominal ultrasound is performed which confirms pyloric stenosis and he was admitted for medical stabilization prior to surgical correction. A nasogastric tube and peripheral IV were placed. He has since been adequately fluid resuscitated and now has good urine output; electrolytes and vital signs are within normal limits.

Questions:
Preoperative Preparation:
1. What other information would you like regarding this patient?
2. What are your initial thoughts regarding the anesthetic plan?
3. How would your plan change if the patient did not have Pierre Robin? What if the patient had another condition other than pyloric stenosis?
4. What complications are you concerned about?
5. What airway equipment would you like to have available? Drugs? Adjunct personnel?

Intraoperative:
1. Any premedication?
2. Do you routinely suction through an NG tube? Perform gastric lavage? Would you use the 10F tube in-situ or place a larger bore tube?
3. Would anyone consider an “awake look”?
4. What induction technique would you use? What if the IV infiltrates?
5. How about a “sedated look”?
6. What if the patient desaturates during airway manipulation?
7. What techniques are available to improve your fiberoptic laryngoscope view?
8. After the ETT is secured, the patient is repositioned and a large leak is heard, end-tidal carbon dioxide is decreasing? What are your thoughts?
9. What is the plan for maintenance of anesthesia? Would this include narcotics?
10. How would you extubate this patient?
11. What concerns do you have regarding recovery?
Discussion:

Alone, the conditions of Pierre Robin sequence and pyloric stenosis present challenges to the anesthesiologist. Together, the two conditions present the conundrum of safely managing an expected difficult airway in an uncooperative patient with a full stomach.

Pathophysiology

Pyloric stenosis is one of the more common infant surgical conditions\(^1\) with an incidence of approximately 1/400 live births and a preponderance for firstborn males. It presents clinically with persistent and frequently projectile nonbilious vomiting, visible peristalsis, dehydration and in extreme cases, hypovolemia. Classically associated are hypochloremic hypokalemic metabolic alkalosis and compensatory respiratory acidosis. Infants are typically between 2 and 8 weeks of age at the time of presentation\(^2\); diagnosis is made by both history and the clinical finding of an olive-sized mass in the epigastrium. Contemporary diagnosis is facilitated by ultrasonography showing thickening of the pylorus. Pathologically, pyloric stenosis is characterized by gross thickening of the circular muscle of the pylorus leading to gradual obstruction of the gastric outlet. Hypovolemia and electrolyte disturbances must be corrected prior to proceeding with anesthesia and surgical treatment. Infants with pyloric stenosis are considered to be at risk for aspiration secondary to their gastric outlet obstruction in addition to the other recognized risks associated with anesthesia for this age group.

Pierre Robin sequence was first described in 1923 by Pierre Robin, a French ‘stomatologist’ and it is estimated to affect 1/8500 live births. It is characterized by micrognathia, glossoptosis, and U-shaped cleft palate. Pierre Robin sequence can be found in isolation or in association with other congenital anomalies. The underlying abnormality is thought to be hypoplasia of the mandible prior to 9 weeks gestation that leads to displacement of the tongue posteriorly and superiorly between the palatal shelves preventing their fusion. Individuals with this defect are known to have difficult airways and often experience airway obstruction, especially in the supine position. The airway is believed to become easier to manage with increasing age.

Clinical

Pierre Robin sequence presents many potential challenges of managing a difficult airway in an infant. Often, the larynx is not visible by direct laryngoscopy, and attempts at awake laryngoscopy are rendered especially difficult in an awake, moving, crying and gagging infant. Unsuccessful attempts at awake direct laryngoscopy may lead to intraoral bleeding that complicates other approaches to securing the airway, such as fiberoptic bronchoscopy. Affected infants often present with upper airway obstruction and encounter difficulties maintaining adequate ventilation and oxygenation in the supine position, the position of choice for performing intubation under most circumstances. Depending on the degree of micrognathia, adequate bag-mask ventilation may be difficult secondary to poor mask seal. A large cleft palate may render visualization of the larynx
and/or navigation of the fiberoptic bronchoscope more difficult. Even after successful passage of the pre-loaded scope into the glottis, there may be difficulty in passing an endotracheal tube over the scope due to both the highly flexible nature of the scope and to possible obstruction by the arytenoids. Infants may be particularly at risk of soft tissue trauma, bradycardia secondary to vagal stimulation, breath-holding, and laryngospasm. Given the infant’s decreased functional residual capacity and increased metabolic demands, such patients are unlikely to tolerate even a short period of breath-holding or laryngospasm without significant hypoxemia.

Alternately, one could consider an awake, but sedated intubation attempt. The goal in this setting would be to provide sufficient sedation to optimize patient cooperation and decrease movement, while maintaining spontaneous ventilation and protective airway reflexes. There are many choices of anesthetic technique combinations that might be applicable, each with its own risks and benefits. More traditional sedative and anesthetic agents include benzodiazepines, opioids, volatile agents and propofol. Many of these agents carry a risk of respiratory depression and blunting of airway reflexes, effects which could be potentially dangerous in the setting of a difficult airway and a full stomach. It should be remembered that both opioids and benzodiazepines have pharmacologic antagonists capable of counteracting respiratory depression and over sedation.

Ketamine is another possibility having the advantage of causing minimal respiratory depression by maintaining near-normal ventilatory response to carbon dioxide. Its use as the sole agent for sedation in the management of the difficult pediatric airway has been described. Ketamine also causes bronchodilation, potentially offsetting intubation-induced bronchospasm. Ketamine use is associated with increased airway secretions which can be countered by antisialogogue prophylaxis.

A relatively novel drug to consider is dexmedetomidine, a selective alpha 2 agonist that has anxiolytic, moderate analgesic, and antisialogogue properties with minimal respiratory impairment; side effects include hypotension and bradycardia. Dexmedetomidine has been successfully used in adult patients as the sole sedative (in combination with local anesthetic topicalization) for fiberoptic intubation. Its use in the pediatric population has also been described (particularly for procedural sedation) and in combination with ketamine in managing a child’s difficult airway. An additional approach is the use of a laryngeal mask in an awake or sedated infant as a conduit for passage of a fiberoptic bronchoscope.

In summary, we describe the conflicting (and substantial) clinical challenges encountered when managing an infant with concurrent diagnoses of pyloric stenosis and Pierre Robin sequence. While many possible pharmacological and technical approaches to these challenges exist, thorough preoperative evaluation and preparation along with immediate availability of capable assistance are the common denominators to successful and safe management.
References


