A case of a neonate with a congenital laryngeal web: management of a difficult airway and intra-operative complications

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Goals:

1. Discuss the differential diagnosis of stridor, aphonia and abnormal cry in a newborn.
2. Review airway management in a neonate, particularly when this is a collaboration with ENT residents and faculty.
3. Debate communication styles and conflict resolution strategies when there is disagreement between two faculty over the need for pre-planning
4. Describe the risks and complications associated with rigid bronchoscopy, emergent tracheostomy, jet ventilation.
5. Discuss critical incident management and effective communication with the family

Case description:

A 3 kg infant born at term is noted to have intermittent stridor and an aphonic cry shortly after delivery. ENT performs a bedside flexible laryngoscopy and makes a preliminary diagnosis of possible laryngeal web. The patient is scheduled for diagnostic laryngoscopy and rigid bronchoscopy with possible resection in the OR.

Questions:

What is a laryngeal web? What are your concerns? Are congenital laryngeal webs associated with any other anomalies?
What are other causes of abnormal cry and stridor in newborns?
Are there pre-operative tests or labs that you would order before rigid bronchoscopy?

Preoperative Evaluation, Chart review and Preparation

Vital signs are age appropriate. Examination of the infant is significant for an aphonic cry. Lungs are clear to auscultation, no retractions. Chest x-ray demonstrates normal lung fields. Echocardiogram is normal. Team has patient scheduled for a

possible chest CT or MRI, which ENT feels can be deferred for now. You approach the ENT physician about formulating a plan for the OR, only to be brushed off and told "we'll see as we go".

Questions:
Do you require additional tests? When might you first want a spiral CT?
What would your plan be for a safe induction?
Would you perform an awake intubation? What is the data for this?
How would you maintain anesthesia?
What would you have available for emergency equipment and monitoring?
What are your options for collaborative airway management and ventilation during this case? Which would be the safest for this patient?
What are the pros and complication risks of jet ventilation?
Would you consider the use of NIRS monitoring for this case?
What are some strategies that could be utilized to improve communication with the surgery team?

Intraoperative management

In the operating room, mask induction is uneventful, an IV is placed, the anesthetic adjusted, and the patient remains spontaneously breathing for initial visualization by surgeon. Following application of topical lidocaine, direct laryngoscopy with rigid bronchoscopy confirms the pre-operative diagnosis of a congenital laryngeal web. ENT is debating how to proceed because the web appears thick, and the airway subglottic size narrow. To assess further, surgeon requests micro- suspension and jet ventilation. You provide same and after 15 minutes of deliberation, the surgeon determines that the thickened nature of the web and narrow airway dictate a secure tracheostomy airway, with delayed web resection and possible tracheal reconstruction. The patient is intubated with a 2.0 ETT under direct visualization. Placement through vocal cords is confirmed visually by the surgeon and yourself and positive pressure ventilation initiated. ETT has a small leak at 15 – chest rise is minimal. End-tidal CO2 is present but there is a poor waveform; breath sounds are faint bilaterally. Oxygen saturations have dropped from 100 to 80% and BP and HR are stable .

Questions:
What is your differential diagnosis for this desaturation?
What are your next steps?
Would you give any additional medications at this time?

Intraoperative management, continued

The decision is to call for a chest xray, but your surgeon wants to move forward with the tracheostomy until the tech arrives. You position, and proceed. Two minutes into the tracheostomy, the patient's heart rate declines first to 90 then to 45, blood

pressure by cuff is undetectable, although pulse oximetry is working and and oxygen saturations are 50%. You auscultate again and hear very faint breath sounds on the left and absent breath sounds on the right. The surgeon suggests you might be mainstem intubated.

Questions:
Differential diagnosis? What is your next step?
What makes a tracheostomy more difficult in a newborn?
Is continuing with an emergent tracheostomy the best plan?

Intraoperative management, continued:

The surgeon completes the tracheostomy quickly, while the anesthesia team delivers epinephrine, performs chest compressions and switches to ventilation through the tracheostomy The heart rate, blood pressure and oxygen saturations improve: HR is 160, pulse ox 88% on 100% oxygen; but the patient continues to appear mottled. Xray tech finally arrives -the chest xray demonstrates a large left pneu mothorax and small right pneumothorax.

Questions:
When would you perform a needle thoracentesis vs a primary chest tube insertion?
How would you perform a needle decompression?
Why do you think this patient developed a pneumothorax ?
Would you place any other monitors at this time?
Who performs neonatal chest tube insertions?
What would you tell the family regarding outcomes after CPR in the operating room?

Postoperative management:

Right side needle decompression is performed in the operating room with significant improvement in oxygen saturation. Bilateral chest tubes are placed. The patient remains hemodynamically stable with adequate ventilation and you prepare for transportation to the NICU.

Questions:
What would be your post-operative plan for sedation?
Would you paralyze? Pros and cons?

Discussion:

Neonatal airway lesions can lead to significant airway compromise and are best approached with a collaborative team effort between the anesthesiologist, neonatal intensive care unit, and surgeon. Congenital laryngeal webs oftentimes present in infancy with aphonia, though when less severe may be an incidental finding on intubation later in life. The incidence of a congenital laryngeal web is 1 in 10,000. Congenital laryngeal webs have been associated with cardiovascular abnormalities, chromosome 22q11 deletion, and velocardiofacial syndrome. The severity of symptoms is related to the degree of airway obstruction and grade of webbing.

Stridor is a common presentation of laryngeal obstruction. Supraglottic or glottic obstruction will usually present as inspiratory stridor, biphasic stridor normally represents a subglottic obstruction, and expiratory stridor often represents obstruction in the distal trachea or main bronchi. Laryngomalacia is the leading cause of stridor in the neonate and accounts for 65-70% of congenital laryngeal anomalies. Vocal cord immobility is the second most common congenital laryngeal disorder, and accounts for 10-20% of anomalies. These infants often present with a weak, breathy cry, or if bilateral can present with biphasic stridor and a preserved cry. Subglottic hemangiomas account for approximately 1.5% of laryngeal abnormalities and may present with recurrent croup, biphasic stridor and cutaneous hemangiomas. Other more rare laryngeal anomalies include laryngeal atresia, congenital subglottic stenosis, laryngeal cysts, saccular cysts, vallecular cysts, and thyroglossal duct cysts. When laryngoscopy fails to establish a diagnosis, further imaging such as MRI or spiral CT may be helpful to delineate lesions that may case external compression of the airway. These lesions may include vascular rings, retropharyngeal masses, localized inflammation or mediastinal masses. Stridor in the infant with a vascular ring, such as double aortic arch, is often worse with feeding and commonly presents after 6 months of life. Some centers approach stridor first with a CT, as a bronchoscopy in the face of a hemangioma you are unprepared to treat, or an airway polyp that becomes swollen or dislodged can be lethal. In this case, the laryngeal web was strongly suspected based upon the preoperative flex laryngoscopy.

Induction of anesthesia in a neonate with an obstructive lesion such as a laryngeal web usually involves an inhalation induction with maintenance of spontaneous ventilation. This is more difficult to accomplish in the neonate as immature respiratory drive and decreased FRC predisposes the infant to hypoventilation and hypercapnia. Maintenance of anesthesia will depend on the severity of the lesion, and may include a total intravenous anesthesia technique or volatile anesthetic. Most practitioners utilize topical agents such as lidocaine. This will reduce the MAC requirements for the rigid evaluation. In a neonate, this may be particularly important as the reported MAC needed without a topical agent approaches 2.5 MAC. In our experience, 2 MAC of sevoforane in a neonate may precipitate apnea and/or hypotension.

Airway management during neonatal endolaryngeotracheal surgery is a challenge due to the variability in location of the surgical site as well as the narrowness of the neonatal airway. Options for management of the airway during a resection include jet ventilation, spontaneous ventilation with or without intermittent intubation, and apneic anesthesia.

Each of these techniques has inherent risk, though jet ventilation is becoming increasingly popular as it facilitates improved operative conditions. Complications of jet ventilation include pneumothorax, drying of the laryngeotracheal membranes, CO2 retention, barotrauma, and pneumomediastinum. In this patient with a very stenotic airway, it is also important to consider the effects of positive pressure ventilation and over-distention of the alveoli due to the need for prolonged expiratory time and auto-PEEP.

We have elected to present to you a pneumothorax complication that was not immediately evident until evolution to a tension pneumothorax. With the initial desaturation event the additional differential to consider includes a second anomaly, a kink in the endotracheal tube, secretions causing plugging, ETT malposition , accumulated atelectasis and underventilation. Proceeding with the tracheostomy at this time is controversial. In many centers the time lag in obtaining a stat chest xray makes this impractical if one suspects a progressive pneumothorax. Needle thoracentesis is indicated if a tension pneumothorax is suspected. To perform needle thoracentesis a syringe is attached to a 14-16G iv catheter, and advanced into the 2nd intercostal space, mid-clavicular line, aspirating continuously. Aspiration of air will confirm correct placement in the pleural space. NIRS monitoring can be helpful in assessing for a tension pneumothorax, as when cardiac output is compromised the NIRS will drop in excess of the arterial desaturation. In addition NIRS algorithms for changes have a shorter time lag than standard pulse oximetry.

Postoperative paralysis is probably not necessary. Additionally, obtaining neurologic assessment is beneficial. In many neonates, modest dose opioids or dexmedetomidine will provide appropriate sedation. Outcomes data would suggest that brief intraoperative CPR has good outcomes. When critical incidents occur, they must be discussed with parents. Some components of this disclosure include the presence of both physicians involved in the care, speaking with the family after they have seen their child settled in the NICU, allowing for questions, avoid any blaming , and following up serially.

Additional Preoperative Decision Making-and Team planning

Team communication and planning is critical in shared airway cases. Making up the plan as you go is sub-optimal, and does not allow for adequate preparation. When communication is inadequate, there are a few key points to improving communication. Speaking to your colleague privately minimizes the conflict aspect. Use of “I messaging” as in “I am uncomfortable proceeding….I have some questions before we proceed” is one approach. Acknowledge that the final procedure may be wait and see, but that all possible options should be laid out to ensure all supplies are ready. Have your questions set for your colleague (Is there a chance of Laser resection? Jet ventilation? Spontaneous ventilation vs. paralysis? Under what conditions might you need to trach? What has family been told in terms of risks and outcomes postop? Is an ICU bed scheduled?) Proceeding to the operating room and then finding out about any of this at the “time out” -post induction- is too late in an airway case!

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