Chapter 15
Head and Neck

Robert S. Holzman

An 8-year-old girl with Klippel-Feil syndrome, scoliosis, a solitary kidney, neurogenic bladder, sacral agenesis, and tethered cord was scheduled to undergo cervical spine fusion following 1 month in halo traction. She was on no medication and developmentally was apparently doing well. She had a known difficult airway from multiple prior surgeries.

R.S. Holzman, MD, MA (Hon.), FAAP
Senior Associate in Perioperative Anesthesia, Boston Children's Hospital, Boston, MA, USA

Professor of Anaesthesia, Harvard Medical School, Boston, MA, USA
e-mail: robert.holzman@childrens.harvard.edu

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Preoperative Evaluation

Questions

1. Will this patient have a "difficult airway?" What are the contributing reasons that you would be concerned about? Why has she been in traction? Is it likely that she has to remain in traction, or can this be released for induction? Should the halo device be removed, or should it remain? Are you surprised by the patient's comorbidities? What developmental explanation can you offer?

2. Does this patient require further evaluation or consultation, and if so, with whom? Any other studies you would like?

Preoperative Evaluation

Answers

1. This patient is highly likely to have a difficult airway for several reasons. First of all, she has Klippel-Feil syndrome as a result of fusion of several cervical vertebrae. Klippel-Feil syndrome severity is typically classified as type I, when patients have a single-level fusion; type II, when patients have multiple, noncontiguous fused elements; and type III, with multiple, contiguous fused segments. It may also be associated with branchial arch anomalies like Goldenhar syndrome, fetal alcohol syndrome, and anomalies of the extremities. Preoperative traction has been known to relieve sensorimotor impairment and improve the quality of the fusion repair. Depending on the type of halo fixation device, it may very well be released or portions released in order to get to the face and the patient's head held carefully in traction by the spine surgeon. The halo in all likelihood will be replaced by Gardner-Wells tongs or a similar device for the procedure. The comorbidities of renal anomalies often occur with Klippel-Feil syndrome; hearing impairment is not uncommon as well. Sacral agenesis and a tethered cord are not typically associated, even though they represent neural crest migration defects.

2. It would be advisable to consult with the ORL service in case their help is needed for securing the airway or for emergency tracheotomy. Soft tissue imaging studies, such as a soft tissue x-ray of the neck, would help define the anatomic relationships, especially the relationship of the posterior pharyngeal wall, which is likely to have moved anteriorly, thus narrowing the pharyngeal cross-sectional diameter and the larynx. This is also an important consideration for the patient's airway status postoperatively.

Intraoperative Course

Questions

1. What monitors will you choose? Why? Does this patient need an arterial line? Why? Does this procedure require any other special monitors? Would a precordial Doppler be a reasonable choice? Why or why not?

2. What are your considerations for anesthetic induction? Your colleague stops by and suggests an awake intubation? What do you think? You select an intramuscular preinduction technique in the pre-op holding area with ketamine because of the extreme separation anxiety, and the patient obstructs within 30 s in the mother's arms and begins to turn blue. What do you do next? Will an oral airway help? Is this patient a difficult intubation? Should you continue with the case?

Intraoperative Course

Answers

1. Routine noninvasive monitoring plus an arterial line to follow mean arterial pressure. In addition, there is a chance of air embolism because as bone is decorticated, the potential point of air entry is superior to the venous system and the right atrium of the heart. The Doppler should be placed in the typical position on the anterior chest wall, but should not be allowed to compress the skin with undue pressure.

2. Depending on the airway assessment and the patient, anesthetic induction might precede intubation of the trachea or follow intubation. An "awake" (actually, sedated, with topical and/ or local anesthetic) can be accomplished in almost any age patient with adequate planning and time. Topical lidocaine can be nebulized or gargled, specific nerve blocks for the glossopharyngeal and superior laryngeal nerves can be administered, a transtracheal injection of lidocaine can be delivered, and intravenous sedation can supplement the entire procedure. At that point, either a direct laryngoscopy, a video laryngoscopy, or a fiber-optic intubation (transnasal or transoral) can be accomplished. The patient can also undergo an inhalation induction with preservation of spontaneous breathing. Neuromuscular blockade can also be utilized once the ability to ventilate by mask is ensured. All of these possibilities are ultimately determined by the anticipated difficulty of the airway as well as the maturity, cooperation, and willingness of the patient and family.

   Sedation or preinduction strategies in the pre-op area have to be judiciously weighed against the risks of proceeding to the operating room, where more familiar emergency surroundings and equipment are available in case of an emergency. The first intervention with acute airway obstruction should be mask positive-pressure ventilation. It may be possible to insert a laryngeal mask airway, but this may also be difficult if the approximation of the laryngeal inlet and the posterior pharyngeal wall is very close, as a result of the traction. This may also make the insertion of an oral airway difficult, although a nasopharyngeal airway might be easier and more effective. Desperate situations almost always evoke some measure of desperate interventions, but if mask ventilation can be provided, then the patient should be moved to the operating room directly where the full range of definitive treatment can be provided.

   The patient is likely a difficult airway because of the underlying anatomic problem and also because of the halo device bringing the posterior pharyngeal wall into approximation with the laryngeal inlet. In addition, exposure of the larynx with a standard laryngoscope may prove challenging in the patient in a halo.

3. What will be your primary anesthetic technique? Why? Will you use nitrous oxide? Why/why not? What is your choice of muscle relaxant, if any? Why? What would you consider optimal fluid management for the case? Choice of fluid? Volume?

4. During surgery, the patient suddenly develops a drop in blood pressure, bradycardia, and a drop in end-tidal CO_2. What is your differential diagnosis? How can you go about narrowing the possibilities? What would you do? Why? Blood pressure is 60/40 with a heart rate of 60; how would you manage his depth of anesthesia? Why?

If adequate ventilation is ensured and the patient's airway is secured, then the team should discuss whether to proceed or not. Reasons not to proceed would include, for example, undue manipulation of the head and neck and the desire to wake the patient up and reevaluate the patient's neurological status.

3. The main anesthetic consideration intraoperatively is choosing a technique that will interfere least with measurement of sensory and motor evoked potentials, so the inhalation agent use has to be minimal, neuromuscular blocking drugs should be avoided, and a hypnotic and narcotic technique would probably be optimal. That combination might typically include propofol and fentanyl, sufentanil, or remifentanil by infusion. While the intraoperative goal would be minimal interference with evoked potential monitoring, emergence and perioperative goals would be directed to comfort as well as ease of assessment of neurological status, which is another reason that narcotic techniques are favored. Minimizing intravenous fluid administration would decrease the amount of tissue edema in dissected areas, which is particularly concerning neural tissue. The choice of fluid should be normal saline rather than lactated Ringer's solution because of the hypo-osmolality of Ringer's, and the volume administered should be conservative.

4. Several causes can be speculated. Sudden volume loss from surgical bleeding is a possibility, and because of gravity, blood loss will not necessarily be visible and obvious. A venous air embolism can produce an identical clinical scenario, as can reflex-mediated neural output from the brainstem while dissecting in the upper cervical spine. While the precordial Doppler is very sensitive for detecting air, it is not infallible, so the surgeon should be informed, the field flooded, and other causes ruled in or out. In that context, the surgeon should be immediately informed of the patient's instability and flood the field. Release of traction will relieve extrinsic pressure from the brainstem if this is a contributing factor. In any event, the patient's depth of anesthesia should be decreased in an effort to ultimately increase regional blood flow and perfusion pressure.

Postoperative Course

Question

1. Would you leave this patient intubated? Why/why not? What criteria will you use to decide about extubation? Why? What technique would you use for extubation? Is it likely that it will be a "difficult extubation?" What would you look for at the end of surgery to help you predict the success of your extubation strategy? How could you use a tube exchanger to facilitate extubation? At the end of the procedure, following extubation, desaturation recurs while the patient is struggling, bearing down, and breath holding. Your management? Why? Is it likely that the patient's airway will be worse after surgery than before?

Additional Questions

Questions

1. A 1-year-old, 9 kg boy is scheduled for repair of orbital hypertelorism for Apert's syndrome. He appears congested but afebrile, with a blood pressure of 92/55 mmHg, pulse 120 bpm, respiration 32/min, and temperature 37 °C. Hematocrit is 33 %. He is on no medications, and his parents report that he is terribly afraid of doctors. Will this patient have a difficult airway? What monitors will you select for this case? What are your considerations for induction of anesthesia? Any anticipated difficulties with vascular access?

Postoperative Course

Answer

1. Postoperative intubation may be a very reasonable choice if the airway was significantly altered by the surgical procedure, as it often is, because of a shortened anteroposterior pharyngeal diameter.

   Edema may be a further consideration following the prone position and significant fluid exchange. But leaving the trachea intubated has relatively little to do with waking the patient up enough to obtain a neurological exam, which should be easily achievable in order to evaluate the neurological status. Another possibility is extubation over a tube exchanger, in order to facilitate reintubation, and this should be considered as well. The mental status does help in assessing fitness (and motivation) for extubation.

   Tube exchangers are a means of providing a stent with oxygen flow to a patient, as well as facilitating reintubation. They are fitted with a locking 15 mm OD circuit connector at the end and can be inserted easily.

   The patient can be struggling, bearing down, and breath holding because he is light and was intubated prematurely or because of secretions or airway irritability.

   The patient's airway sill likely be worse – more narrowed – immediately after surgery, but will remodel itself as an adaptive measure.

Additional Questions

Answers

1. That depends on what you mean by a difficult airway. The midface is hypoplastic and the eyes appear to be proptotic, although that is more a reflection of the hypoplastic midface and orbits but normal-size eyes. The hypoplastic skull base contributes to abnormal development of the sphenoid, frontal, and maxillary sinuses, which in turn often leads to an appearance of chronic congestion, simply because sinus and nasal drainage is impaired. The branchial arches, however, are typically not affected, so that mandibular development proceeds normally. The combination often results in chronic upper airway congestion, moderately difficult fit for a mask, but relatively easy laryngoscopy and intubation. The surgical approach is a bifrontal craniotomy with multiple osteotomies and (hopefully) preservation of an intact dura. There can, however, be neurosurgical consequences if there is a dural puncture or even with the prolonged reconstructive surgery, dural exposure, and large blood loss and fluid shifting that occurs. Polysyndactyly of the hands and feet is common, and therefore, intravenous access may be difficult.

2. What is Goldenhar's syndrome? How does it develop? What are the anesthetic considerations? Are there particularly significant associated anomalies? Do these patients tend to get easier to take care of over time?

3. A 5-year-old was hit by a car while riding his bike and sustained an over-the-handlebar fall into the pavement, suffering a Le Forte I fracture, mandibular fracture, a Colle fracture, and various bumps and bruises. He comes to the operating room in a soft collar in preparation for oral surgery to repair his facial trauma. What is the difference between the various Le Forte fractures, and what is the significance for your anesthetic plan? Your considerations for anesthetic induction? Maintenance? What are your considerations for anesthetic emergence? What are your considerations for how you will wake him up? Would it be safer to leave him intubated? Why or why not?

Considerations for anesthetic induction include the patient and family's emotional state – the patient is fearful because of multiple visits to the clinic as well as "stranger anxiety" typical for this age, and the parents are fearful as well because they know this is a big operation with significant morbidity. There is also the larger context of the uncertainties of the family with a chronically ill, syndromic child. While there may be advantages to a parent present during induction in the operating room such as decreased crying and decreased aerophagia, thereby lowering the risk of intragastric air and regurgitation, these advantages may be outweighed by the relatively minimal chance that this infant will be consoled by a parent's presence, the likely difficulty with the mask fit because of midfacial hypoplasia, and the presence of impaired secretion elimination because of the sinus abnormalities with the strong possibility of airway irritability, laryngospasm, or bronchospasm on induction.

A balanced technique with fentanyl/air/oxygen and isoflurane would be my anesthetic technique of choice. Nitrous oxide should be avoided because of the risk of venous air embolism and also because body cavities will be opened and subsequently closed. Muscle relaxation should be utilized to ensure lack of movement.

2. Goldenhar's syndrome is the eponym for hemifacial microsomia, an anomaly characterized by variable hypoplasia of the mandibular division of the first branchial arch, including the mandibular ramus, body and temporomandibular joint, hypoplasia of soft tissue components of the face and jaw, and hypoplasia of the facial nerve. The more severe forms are typically very difficult intubations because of the inability to open the jaw on the affected side. Furthermore, breathing through a natural airway (and therefore support through a mask airway) may be difficult because of a small pharynx. Because this anomaly occurs early in embryological life, anomalies of other contiguous areas are not uncommon, including fusion of occipital somites that can give rise to the Klippel-Feil anomaly of cervical vertebral fusion. Cardiac defects such as atrial and ventricular septal defects may also occur. A paramedian cleft palate may also occur. Anomalies of the first branchial cleft such as low set, misshapen ears, and sensorineural hearing loss are common as well. Managing the airway of these patients tends to get more difficult with time.

3. You have to be concerned as much about what you don't see as what you do see; the fact that he has multiple facial fractures and therefore the potential for airway trauma or difficult laryngoscopy and intubation may be the tip of the iceberg; he may also have chest and cardiac contusions, blunt abdominal trauma, closed head injury, and a period of unconsciousness and altered sensorium with or without concussion. All of these are less obvious in physical exam and therefore must be suspected and inquired about during the history, with any additional appropriate laboratory tests. Assuming that these other issues have been ruled out, then the amount of bleeding, soft tissue injury with swelling, bony injury, and trismus will affect the ease of induction, mask fit, and intubation. A 5-year-old will not

4. What is the developmental history of a cleft lip and palate? What associated anomalies should you expect? Are there other associations of cleft palate with craniofacial deformities? How can you assess the potential for intubation difficulty? Would you use a muscle relaxant as part of your anesthetic technique? Narcotics? Why does a cleft palate develop with Pierre Robin sequence?

5. A 9-month-old is scheduled for exploration of the posterior triangle of the neck for a large right-sided “cystic hygroma” that is half the size of his face. What is a cystic hygroma? From where does it arise? Why does the surgeon want to do a posterior triangle exploration? What are your anesthetic considerations? Are these patients difficult to intubate? Under what circumstances? He asks you to leave the right arm free of IVs in case he has to do an axillary exploration. Why would he ask that?

likely tolerate an awake laryngoscopy and intubation, so a decision will need to be made about the amount of trismus present that is likely to be relieved with the induction of anesthesia (and therefore facilitate intubation) versus the amount of soft tissue swelling and/or bony injury that may make laryngoscopy and exposure difficult. Radiological evaluation may be very helpful in this regard, especially if stridor is present. It is important to discuss the extent of the repair, as it is likely that a nasal intubation will be necessary if internal maxillary fixation is to be applied. Emergence considerations are important as well; coughing, bucking, and Valsalva maneuvers should be avoided, and in general, a smooth wake-up is best. It may be well advised to keep the patient intubated for several days until soft tissue swelling decreases and an air leak is reestablished. Extubation may be accomplished in the intensive care unit or in the operating room.

4. Typical cleft lip formation occurs along a line joining the primary and secondary palates through the middle of the ipsilateral ala. Clefting of the primary and/or secondary palate may also occur in continuity with a cleft lip. Clefting of the soft palate may also occur, because palatal fusion occurs progressively from anterior to posterior portions of the palate. There may be associated anomalies of structures or organ systems that are developing at the same time as the midface and palate, such as the heart and occipital somites. A cleft palate may also occur in association with glossoptosis (Pierre Robin sequence), in which case it is more properly considered a deformation by interference with the progressive midline fusion of the maxillary shelves. A paramedian palatal cleft can occur with branchial arch abnormalities such as hemifacial microsomia (Goldenhar's syndrome) [3]. It is important to carefully evaluate any association with branchial arch abnormalities because of the potential for difficult laryngoscopy and intubation. The use of any specific anesthetic technique should be directed to the desired endpoint of the surgery as well as any anticipated difficulty with extubation. These patients will occasionally have significant obstructive or mixed sleep apnea; attention to perioperative monitoring for apnea and hypoxia is probably more important than any particular anesthetic technique.

5. "Cystic hygromas," more properly known as cystic lymphangiomas or macrocystic lymphatic malformations, typically present in this area of the head and neck as lobular, multiloculated collections of cysts and lymphatic vessels filled with clear or serous fluid. There are a variety of often bewildering and challenging presentations because, while most arise in the posterior triangle of the neck in continuity with the fascia of the longus colli muscle, they may extend into the axilla, the anterior triangle of the neck, and intraorally. In addition, bleeding may occur into the cysts, forming clot and therefore hard, extrinsically compressive masses to the oral cavity and therefore the airway. The airway may therefore be difficult to visualize in these patients, and each should be looked at individually for the potential for difficult mask fit, visualization, and tracheal intubation.

Specific and General References

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