Difficult Airway in a Child with Epidermolysis Bullosa who presents for PEG-tube and MRI

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Objectives:
- Discuss preoperative evaluation, preparation, disease progression and anesthetic implications associated Epidermolysis Bullosa (EB).
- Review pathophysiology of EB and Identify suitable sedative and anesthetic choices for a patient with EB presenting for non-invasive and invasive procedures.
- Devise a strategy for managing the challenges related to airway management in patients with EB.
- Discuss the recognition and management of the difficult airway in off-site locations

Case History:
A 15 year-old, 40 kg female with epidermolysis bullosa (EB) presented for a PEG tube and MRI. She has suffered from progressive dysphagia and aversion to food prompting this procedure. Her arms are wrapped in dressing and she has skin blisters of varying degrees all over her body, including her face.

Questions:
What is Epidermolysis Bullosa? Are there different types of this disease and is there a spectrum of severity? Describe the course of this disease. Does it get worse with age? What are the anesthetic implications of EB? What is the usual life expectancy of these patients?

Case history and physical examination (continued):
Her EB is recessive dystrophic epidermolysis bullosa and her face is riddled with blisters, especially around her mouth and lips. Her dressings are changed 3 times daily by her mother and she is wrapped in blood-tinged burn dressings from her neck to her feet. Her malnutrition
is progressing to chronic weakness and decreased ability to perform routine tasks of daily living. Her medication includes Zoloft and Xanax for anxiety and depression. Her last anesthetic was for an esophageal dilatation and her family has no information regarding the anesthetic management at that time. Physical exam revealed normal respiratory and cardiac examination. She is very thin, appears tired and has significantly reduced mouth opening. Her room air oxygen saturation is 98%, and all other vitals are similarly normal. Her electrocardiograph from 3 years ago reveals prolonged QT interval. She has not had anesthesia for 3 years. Her last anesthetic was for an esophageal dilatation and her family does not believe there were any issues, except the anesthesiologists told them the “breathing tube didn’t take right away.”

Questions:

What is the typical management strategy for someone with EB? Are there other concerns besides the skins lesions? She has a seemingly uncomplicated experience during her last anesthetic. Does this translate to proceeding to the operating room without reservation? If she does in fact have a difficult airway, what is the next step in preparation for this case? (ENT/Pulmonary consult).

Preoperative Studies:

EKG reveals normal sinus rhythm, with a QTc of 503 ms. Echocardiogram from 3 years ago revealed normal right and left heart function mild TR, and otherwise normal. Chest radiograph was clear and unchanged from 3 years ago. Complete Blood Count revealed a mild leukocytosis (12,000), H and H was within normal limits and renal panel revealed only mild hypocalcemia. Of note, her chart was obtained from her previous anesthetic 3 years ago which suggests the anesthesia provider was unable to visualize the glottis secondary to limited mouth opening and extensive erythema and blood in the airway after instrumentation and that they placed an LMA fiberoptically passed an ETT through the LMA.

Questions:

What risks and possible complications should be disclosed to the patient (who is exceedingly nervous) and her parents? Would you mention worsening blisters around the face or body? Would you mention tracheostomy or death as a possible complication?

Mom was wondering what safeguards you will take to prevent worsening of the blisters/trauma to the skin?

The resident in the room has never taken care of an EB patient before and asks how to secure the IV, place a mask on these patients, place EKG leads on this patient, etc? Do you have answers to his questions?
The resident, still nervous, ask how do I touch the patient without hurting her?

**Case Progression:**

After consulting with the pulmonology and Ear/Nose/Throat surgeon, a plan was formulated regarding how to secure the airway for this procedure, if need be. The family was on board with our plan and agreed to proceed.

**Questions:**

What is your anesthetic plan? Local vs. Sedation vs. General Anesthesia? What are the pros/cons of each? How do you plan on establishing IV access in this very nervous teen who has arms and legs extensively bandaged over oozing blisters? Would you perform a nitrous/masked induction in order to establish IV access? Would you use ultrasound? If you used local anesthetic technique, which drugs would you choose? How would you provide oxygen? If you performed MAC, which drugs would you choose? How would you provide oxygen? If you performed general anesthesia, which drugs would you use? Would you paralyze the patient? How would you secure the airway? What are your options? How would you extubate this patient? At what point do you place an LMA? At what point do you consider emergency airway? How painful is this procedure? What drugs would you use for perioperative analgesia? Patient has a history of motion sickness and PONV? Would you give this patient ondansetron? If not, what will you do to prevent PONV? Do you plan on sending this patient home after anesthesia? How long would you keep her in PACU?

**Intraoperative Care:**

The plan was to perform monitored anesthesia care on this patient, with generous amounts of local anesthetic, while maintaining spontaneous respiration. Several attempts were made to place the intravenous in same day surgery, but to no avail. How do you proceed? The patient became increasingly more upset and non-compliant and begged for nitrous oxide for IV placement. What concerns do you have about using a mask on child with EB and blisters around the face? How do you mask this child without producing more trauma to the face? Patient was brought into the operating room and Nitrous oxide was used for ultrasound guided-placement of right antecubital 20 g IV. Then 2mg of midazolam, 1mg/kg of ketamine were given intravenously and a propofol infusion was started at 75mcg/kg/min. A nasal cannula with end tidal CO2 monitoring was placed on the patient and spontaneous respirations were confirmed. After incision, patient began moving. How do you proceed? Fentanyl? Ketamine? More Local? You give the patient 50 mcg of fentanyl and 25 mg of propofol, which deepens the patient adequately. 2 minutes later, you notice no more EtCO2? How would you proceed? Patient becomes exceedingly hypoxic? What do you do next? An LMA was placed, but still could not ventilate patient. How would you proceed? Do you give succinylcholine? Patient is now easier
to mask with 2 hands, however it’s becoming clear this child needs to be intubated for the procedure. How would you proceed? You decide that the patient’s mouth opening is sufficient for a video laryngoscope. However, when you attempt with the C-mac, the patient bucks and you traumatize the oropharynx. The trauma makes successful fiberoptic intubation unlikely? Would you trach this patient? Would you wake the patient up? How would you proceed? Ultimately, you are able to visualize bubbles near the glottic opening using the video laryngoscope and you pass the endotracheal tube successfully. The patient’s airway is secure and the surgeon proceeds with the PEG tube placement.

After successful completion of the PEG tube, Are there any specific concerns you have for this patient, given her difficult airway and EB?

**Postoperative care:**

Decision is made to attempt to extubate patient. What is your extubation plan? Do you take her back to the operating room for extubation? Do you call ENT for extubation? After successful extubation, you realize that the blisters that were on her face pre-operatively are much worse and the trauma in the airway is visible. How do you explain this to the parents? Is this an adverse event? Is this an expected and unfortunate aspect of intraoperative management in patients with EB? What would you do next time to minimize this intraoperative trauma?

The patient recovers uneventfully and is discharged home after she spent one night in the intensive care unit. She returns six months later for an MRI evaluation for recurrent severe headaches. This time, the parents requested you to provide anesthesia for this case since you did a nice job with last procedure.

Questions:

What are the special challenges relating to providing anesthesia in the off-site environment? How and where will you induce anesthesia for this patient? How do prepare for securing the airway in MRI suite? What sedative agent would you consider if you decided to sedate this patient? Would you consider dexmedetomidine (DEX) in this case scenario? What about ketamine? How do you use DEX as a sole sedative in MRI? Why DEX alone or in combination with ketamine could be an attractive choice in this scenario? What adverse hemodynamic affects you should expect when you use DEX as a sole sedative?
Discussion:

Epidermolysis bullosa (EB) is a group of inherited, mechanobullous disorders caused by mutations in various membrane proteins in the skin. There are four major recognized types of EB, depending on the location of the target proteins and level of the blisters: EB Simplex, junctional EB, dystrophic EB, and Kindler syndrome (1).

The prevalence of Epidermolysis Bullosa has been estimated at 1 in 12,000 live births in the USA for mild variants and 1 in 100,000 live births for more severe variants. 2. Epidermolysis Bullosa (2). Between 1986 and 2002, the National Institutes of Health (NIH) funded the National EB Registry (NEBR), a cross-sectional and longitudinal epidemiological study of epidermolysis bullosa patients across the entire continental United States. Nearly 3300 epidermolysis bullosa patients were identified because of scarring, contractures, ankyloglossia, poor dentition, all patients with EB must be considered difficult airways until otherwise proven. Oftentimes the most profound limitation is poor mouth opening in these patients. As such, laryngeal mask airways (LMA) and other airway devices may be difficult to employ. Failed attempts at such measures may traumatize the airway and bleeding may pose even further upper airway difficulties. Even prior successful airway management does not portend a similarly successful outcome in future anesthetics, since the disease progresses and airway management often becomes more cumbersome as the patient ages (3). James and Wark reported 3 cases of intra-oral bullae formation in patients suffering from EB after in 113 oral and 18 nasal intubations, although no immediate postoperative complications were observed (4). Certainly this stresses the need for adaptability in airway management from anesthetic to anesthetic in EB patients.

In addition, almost all patients with EB have GERD from esophageal strictures and are at increased risk for pulmonary aspiration during airway instrumentation and general anesthesia (5).

In summary, the child EB poses many challenges to the anesthesia team. The anesthetic of choice should be individualized depending on the severity and extent of the patient’s symptomology. Airway management, IV placement, analgesia, and finesse with which the anesthetic is conducted necessitate a careful and thought out strategy and even in the best of hands may require improvisation. Ultimately, these patients may present with esophageal strictures, contractures, skin infections; all of which may portend a visit to your operating room. Familiarity with this disease will allow us to be ready for the challenges these children pose.