

# Management of a Difficult Airway in a Child with Epidermolysis Bullosa Dystrophica Using a Combination of Direct Laryngoscopy and Fiberoptic Bronchoscopy

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## Citation

A Rosenbaum, M Follansbee, J Crowley, R Abrams, M Greenberg. *Management of a Difficult Airway in a Child with Epidermolysis Bullosa Dystrophica Using a Combination of Direct Laryngoscopy and Fiberoptic Bronchoscopy*. The Internet Journal of Anesthesiology. 2019 Volume 38 Number 1.

DOI: [10.5580/IJA.54366](https://doi.org/10.5580/IJA.54366)

## Abstract

We describe a novel approach to a difficult airway in a 12-year-old boy with epidermolysis bullosa dystrophica undergoing de-cooing, pseudosyndactyly release, flexion contracture release and full thickness skin grafting of the right hand. A fiberoptic bronchoscope combined with direct laryngoscopic assistance was used to intubate the patient in whom we encountered a difficult airway. By incorporating the fiberoptic bronchoscope, this approach minimized trauma allowing for mucosal examination with placement and removal of the endotracheal tube.

## INTRODUCTION

Epidermolysis bullosa (EB) is a rare disorder characterized by fragility of the epidermis involving the formation of bullae with shearing force ('Nikolsky' sign), as well as ulcers and erosions after relatively minor trauma. These blisters form on the mucocutaneous surfaces due to inherited deficiencies in connective tissue structures (keratin, collagen, laminin). EB is broadly categorized into three subtypes based on histologic patterns: epidermolysis bullosa simplex (EBS), junctional epidermolysis bullosa (JEB), and dystrophic epidermolysis bullosa (DEB). Each sub-type manifests different symptoms with variable life expectancies and surgical needs.

DEB is caused by mutations within the human COL7A1 gene encoding the protein type VII collagen (collagen VII) and can be either autosomal dominant or recessive. DEB is considered the most severe of the survivable forms, with significant scarring and contractures leading to pseudosyndactyly of digits, esophageal strictures, and contractures of the mouth.<sup>1</sup> Gene therapy is in development, and genetically corrected autologous skin transplants have been attempted as a potential cure. Though some success has been shown, palliative surgeries remain a clinical necessity.<sup>2,3</sup> These patients often need multiple surgeries in their childhood, which require special anesthetic considerations

for positioning, monitoring, and airway management given their fragile skin.<sup>4</sup>

Airway management in DEB can be difficult due to microstomia, ankyloglossia, laryngeal stenosis, esophageal strictures, GERD and ease of forming oropharyngeal blisters.<sup>1,5,6</sup> Pre-operative evaluation is especially important in these patients, however, a favorable airway on exam may still pose challenges due to anatomic variation unable to be assessed prior. Careful preparation is necessary, with back up airway devices being available, should direct laryngoscopy be impossible.

Two-hand mask ventilation has been proposed to decrease the sheer force on the facial skin, though long periods of mask ventilation should be avoided. A well lubricated laryngeal mask airway (LMA) has been used with success but there is still a risk of creating oral pharyngeal lesions.<sup>7</sup> There have been a number of reported techniques to intubate the trachea of a child with EB including; traditional direct laryngoscopy, fiberoptic intubation through LMA, blind nasal, awake and asleep nasal fiberoptic, Glidescope® assisted nasal fiberoptic, and tracheostomy.<sup>7,8,9,10,11,12</sup> Depending on the type and duration of the surgery, each of these techniques may be a viable option for intubation and airway management.

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In this report, we present a patient with dystrophic epidermolysis bullosa, in whom tracheal intubation using a fiberoptic bronchoscope (FOB) combined with direct laryngoscopy was used for airway management.

### CASE REPORT

The patient is a 12 year old left handed boy (1.27 m, 22.8 kg) with recessive DEB presenting with recurrent syndactyly and flexion contractures after syndactyly release surgery 5 years prior. He was scheduled for de-cocooning of the right hand with finger flexion contracture release and full thickness skin grafting (Figure 1). Pre-operatively, on examination of the airway the patient was noted to have limited mouth opening, and was a Mallampati II without limitation of neck extension.

#### Figure 1

Right hand prior to de-cocooning, contracture release, and full thickness skin grafting.



Prior to the surgery, a pre-operative huddle was done to familiarize all members of the surgical team to the relevant special needs of an EB patient and to this particular surgery. The patient was then brought to the operating room (OR) after receiving 15mg of oral midazolam to decrease anxiety. The patient transferred himself carefully to the OR table, lying supine upon a well-lubricated gel pad and gel donut headrest avoiding unnecessary pressure on skin or existing bullae. Minimal touch principles were employed while positioning and moving the patient during the case. All adhesive tapes were placed away from the anesthesia workspace and extensive lubrication with Aquaphor™ was utilized on any material in contact with the patient to prevent damage to the skin.

We employed minimal monitoring in an attempt to decrease the risk of skin injury. A lubricated clip-on pulse oximeter was placed on the auricle and a blood pressure cuff was placed over a Vaseline gauze covered lower extremity.

Electrocardiogram monitoring was not used but needle electrodes were available if needed. No esophageal devices were placed, and temperature was monitored with a lubricated probe in the axilla. All remaining exposed areas of skin on the upper and lower extremities were carefully wrapped in Xeroform™ gauze.

Considering the long duration and stimulating nature of the surgery, a general anesthetic with endotracheal tube was planned. A lubricated mask was used for induction, with 70% nitrous oxide and increasing concentrations of sevoflurane. After induction, the eyes were lubricated generously for protection. Special attention was paid while mask ventilating the patient to prevent skin lesions. Intravenous access was obtained with a 22g catheter, which was wrapped with Vaseline gauze and Kling™ rolled gauze, covered with Coban™. Special care was taken to prevent pressure on the skin by the IV tubing and hub (Figure 2).

#### Figure 2

Intravenous access with adhesive free dressing.



The patient was then given 30 mg (1.3 mg/kg) of propofol and 30 mg (1.3 mg/kg) of rocuronium to facilitate endotracheal intubation after mask induction. After paralysis was achieved, direct laryngoscopy with a lubricated Miller 2 blade yielded a grade 4 view and intubation was not attempted. It was determined intubation by direct laryngoscopy was not possible and we decided to switch techniques to include a FOB to assist with visualization of the glottis. One provider performed gentle direct laryngoscopy with a Macintosh 2 blade to open the pharynx while the second provider placed the FOB (Olympus Exera BF-XP160F 2.8mm pediatric fiberoptic bronchoscope), loaded with a 4.5 mm cuffed endotracheal tube, into the oropharynx and through the vocal cords. A few shallow pre-existing ulcers were noted in the oropharynx, and the carina

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was visualized before the ETT was advanced. Tracheal placement was confirmed with end tidal CO<sub>2</sub> and chest rise. Optimal placement was confirmed 2 cm above the carina with the FOB. ETT was secured with a lubricant-soaked twill tie over Mepilex™ to distribute pressure. All points of contact between the ETT security system and the skin were lined with Vaseline gauze (Figure 3).

### Figure 3

Secured endotracheal tube, dressed to avoid direct skin contact and pressure.



Anesthesia was maintained with sevoflurane and a dexmedetomidine infusion titrated from 0.7 to 2.2 mcg/kg/hr based on heart rate and blood pressure. The patient was ventilated on pressure control settings. To reduce post-operative pain and pruritus, an opioid free anesthetic was employed, with nerve blocks, dexmedetomidine, and intravenous acetaminophen 15 mg/kg given at the end of the case. After induction, cuff blood pressures were spaced out to every 15 minutes to minimize pressure and traction on the leg. Vital signs remained stable without need for vasoactive medications or additional monitors.

The surgery proceeded without difficulty with successful pseudosyndactyly release and full thickness skin grafts (harvested from right groin) placed between the web spaces and middle and ring finger proximal interphalangeal joints

(Figure 4). Fingers were fixed in extension with K wires. The groin incision was injected at the start of the case with lidocaine with epinephrine 7mL 1% 1:100000 and the wrist was blocked at the conclusion of the case with 16 ml of 0.25% bupivacaine. The wounds were dressed with Xeroform™ gauze, Kerlix™, and a fiberglass sugar tong cast.

### Figure 4

Right hand post-operatively, with fingers fixed in extension and skin grafts in place.

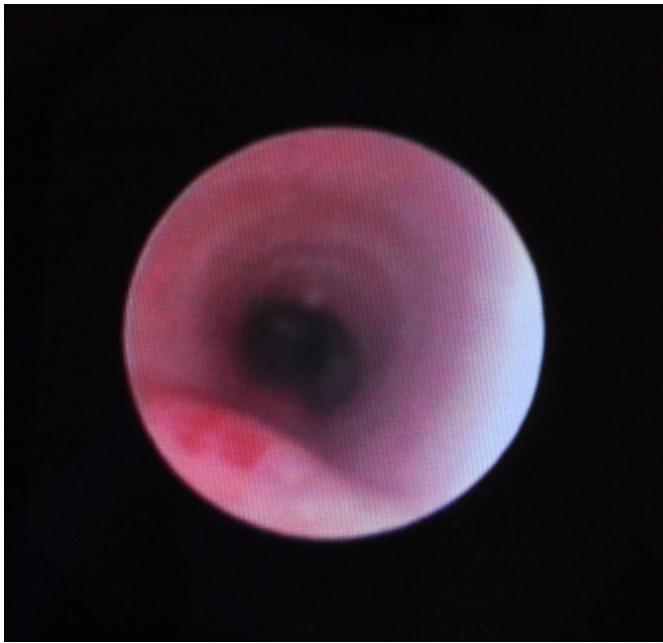


Prior to extubation, all anesthetics were discontinued and the effects of the muscle relaxant were reversed with sugammadex. With the patient still deeply anesthetized, the FOB was passed through the endotracheal tube to visualize the airway structures during extubation. The ETT cuff was then deflated and both the ETT and FOB were carefully removed. With the fiberoptic scope just past the tip of the ETT, the carina, trachea, larynx, and pharynx were assessed for any new trauma or lesions from baseline assessment. One new area of erythema was seen in the posterior trachea at the level of the ETT cuff (Figure 5) and the oropharyngeal ulcers noted on intubation were again observed. No new bullae or ulcers were found on fiberoptic examination of the upper airway. The patient was carefully lifted from the operating table to a gurney using a draw sheet that kept his lubricated gel pads in place. He was transferred to PACU where his recovery was unremarkable, without pain, pruritus, nausea, or coughing. He was stable and discharged home the same day with plan for follow up surgery in two weeks.



**Figure 5**

Bronchoscopic view of trachea upon extubation, showing small area of erythema at level of endotracheal tube cuff.



## DISCUSSION

Patients with epidermolysis bullosa pose a unique challenge to the anesthesiologist, demanding very specific and thoughtful care to avoid injury and airway compromise. Considering that these patients are likely to undergo multiple surgeries in their early years, it is imperative to create a standard of practice, and over the past few decades, there has been a growing body of knowledge supporting the best practices for safe anesthesia.<sup>1,13-15</sup>

Optimal positioning is easily achieved with gel pads, Vaseline gauze, Xeroform™, copious petroleum based lubrication, and minimal touch principles.<sup>14</sup> Monitoring can be streamlined to utilize pulse oximetry for oxygenation and heart rate, non-invasive temperature, and blood pressures cycling at longer intervals to reduce pressure on skin. All adhesive tapes should be avoided, especially in securing the airway and IV access. Less clear is a standard strategy for airway management, as each patient's unique anatomy and surgical factors significantly impact the approach. Many techniques have been reported for patients with EB, each with potential advantages and challenges to be considered while developing an anesthesia plan.<sup>1,7-12,15</sup>

Extreme care must be taken during mask ventilation to avoid undue manipulation, pressure, or shearing forces on the face and mandible. Petroleum lubrication should be generously

applied to the mask and gloves, with minimal mandibular traction, using a two handed technique for ventilation. It is noted that these patients are often easy to mask ventilate due to oral and lingual scarring creating space in the oropharynx without obstruction.<sup>16</sup> While these children may be able to tolerate mask ventilation for short cases (less than one hour), prolonged masking is not ideal, and intubation of the trachea is often necessary for longer duration surgeries. Our patients' procedure took 3.5 hours, and required a completely still surgical field, so we felt intubation was the most appropriate way to manage the airway. Intubation techniques in DEB patients should minimize trauma to the oropharynx and trachea without creation of new bullae or ulcers. Well lubricated LMAs and blind nasal intubation have been used with success, with a low incidence of new bullae formation; however, they are not free of traumatic contact and the lack of direct visualization and inability to assess for new trauma post-operatively makes these methods less desirable

Direct laryngoscopy is the standard for favorable airways with good success in most patients. When a difficult airway is expected, awake fiberoptic intubation may be considered. However, they are not without risk, and are not likely to be tolerated in children. If initial laryngoscopy is unsuccessful, Glidescope®, fiberoptic, or a combination of the two techniques dramatically improves intubating conditions, allows for easy visualization of structures, and can confirm placement of the ETT. The use of a Glidescope® to assist placement of a nasal fiberoptic bronchoscope has been described, with the benefit of having a grade 1 view of the cords while passing the FOB.<sup>12</sup> However, a pediatric Glidescope® is not always available and having to re-instrument with a bulkier glide blade may result in more airway trauma.

Described here is a novel two person oral fiberoptic intubation technique with direct laryngoscopic assistance, after a grade 4 view with traditional direct laryngoscopy. This method may be of benefit to the patient and provider in certain circumstances, especially after a failed initial direct laryngoscopy. Attempting a second laryngoscopy with a different blade is a reasonable choice, which in this case was done with a MAC 2, just before the FOB was inserted. Unfortunately there was no improvement in view on direct laryngoscopy. Simply lifting the laryngoscope already in place to pass the fiberoptic scope was both efficient and minimally traumatic. The MAC blade utilizes the base of

tongue without epiglottal pressure to generate optimal fiberoptic conditions for visualization of the cords. Additionally, the tongue is the most resistant tissue in the oropharynx to bullae formation and can be manipulated carefully without the same concern for epithelial damage as with a Miller blade, a LMA or via nasal intubation. This method may reduce additional damage due to repeated instrumentation of the upper airway, especially if the patient has extensive oral ulceration prior to surgery. In this case, no new ulcers, bullae, or trauma were noted on extubation, except for a small erythematous area at the level of the cuff, unlikely related to direct laryngoscopy or fiberoptic bronchoscopy. The choice of using a small ETT (4.5mm) relative to age of the patient was chosen to lessen the possibility of new lesions from ETT pressure at the glottis.

Anesthesiologists caring for patients with DEB strive to prevent new lesion formation during surgery or as a result of perioperative care. Therefore it is optimal to obtain a baseline examination of the oropharynx with the FOB, allowing image capture for documentation and follow up care. The FOB can also be employed upon extubation to evaluate for anything that may have occurred during the procedure. In this case it was clear upon re-examination that no new trauma was present in the oropharynx, allowing the providers to feel confident that airway management was not responsible for any new oropharyngeal sores. Imaging captured during intubation and extubation may also help with anesthetic planning for future surgeries.

One downside of combining FOB and direct laryngoscopy is that a two-person technique is required. However, since these are specialized cases and should be done in a specialty center it is likely that help would be available. In all but the shortest of cases, the team should plan for a second anesthesia provider to be available during induction in a patient with EB.

## **CONCLUSION**

For patients with epidermolysis bullosa, even with a favorable airway exam, it is prudent to have a fiberoptic bronchoscope available should a difficult airway arise. Simple oral placement of the fiberoptic scope with direct laryngoscopic assistance may be a practical method to secure an airway, avoid extended mask ventilation and minimize oropharyngeal trauma. This technique also has the benefit of

airway visualization, documenting lesions before and after the case, and confirm endotracheal placement prior to the start of surgery.

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