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Anesthetic Challenges in Dystrophic Epidermolysis Bullosa During Upper Gastrointestinal Endoscopy: An Illustrative Case and Narrative Review of the Literature

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Abstract **Case Report**

Dystrophic epidermolysis bullosa is a rare inherited disorder in which cutaneous and mucosal fragility complicates perioperative care. Upper gastrointestinal endoscopy presents particular challenges because airway access is shared and instrumentation can injure fragile tissues. We describe a 20-year-old woman with hereditary dystrophic epidermolysis bullosa referred for esophagogastroduodenoscopy because of persistent dysphagia despite three previous dilation procedures without symptomatic benefit. Contrast imaging demonstrated severe distal esophageal narrowing extending to the cardia. Propofol was administered by target-controlled infusion using the Schnider pharmacokinetic model, titrated to a Modified Observer's Assessment of Alertness/Sedation score of 1–2. Supplemental oxygen was delivered through nasal cannulae incorporating capnography. A videolaryngoscope and flexible bronchoscope were prepared, with tracheotomy equipment immediately available. Recovery was uneventful. A focused narrative review examines airway assessment, aspiration risk, respiratory surveillance, atraumatic monitoring, and tissue protection. This clinical experience illustrates a sedation strategy supported by advance airway-rescue preparation; it does not establish its comparative safety. Perioperative planning should balance the risks of instrumentation against those of an unprotected airway, while maintaining reliable oxygenation, ventilation, and cardiovascular surveillance.

Keywords: dystrophic epidermolysis bullosa; anesthesia; difficult airway; esophageal stricture; gastrointestinal endoscopy; perioperative monitoring.

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INTRODUCTION

Inherited epidermolysis bullosa (EB) comprises genetically heterogeneous disorders characterized by tissue cleavage after minor mechanical trauma. Dystrophic EB (DEB) results from pathogenic COL7A1 variants affecting type VII collagen and anchoring fibrils beneath the basement membrane. Dominant and recessive forms differ substantially in severity; extensive scarring alone cannot establish the inheritance pattern. Current classification incorporates the cleavage level, phenotype, mode of inheritance, and molecular findings [1].

The rarity of EB limits routine clinical exposure. The United States National EB Registry estimated an overall inherited EB prevalence of approximately 11.1 per million inhabitants and an incidence of 19.6 per million live births. These historical estimates concern all inherited EB types rather than DEB alone [2]. For anesthesiologists, the practical importance of rarity is that familiar devices and routine handling may require substantial adaptation.

We use an adult patient referred for upper gastrointestinal endoscopy as a clinical illustration of the challenges generated by the coexistence of esophageal disease, mucocutaneous fragility, and potentially difficult airway access. The purpose is to examine perioperative decision-making rather than to claim the superiority of a particular anesthetic regimen.

CLINICAL ILLUSTRATION

A 20-year-old woman with hereditary dystrophic epidermolysis bullosa was referred for esophagogastroduodenoscopy because of persistent

dysphagia despite three previous esophageal dilation procedures. The clinical phenotype included marked cicatricial deformity of the hands and feet, loss of normal digital separation, perioral changes, and abnormal dentition (Figure 1). These findings prompted attention to both airway access and protection of fragile tissues.

A water-soluble contrast study described a very tight, tapered distal esophageal stenosis extending to the cardia, without a reported esophagobronchial fistula. The radiologist raised reflux-related injury as a possible explanation and recommended clinical correlation. The appearance therefore supports severe structural esophageal disease without independently proving its cause. A thoracoabdominal computed tomography report noted that poor esophageal distension limited assessment and described no clearly identifiable esophageal wall thickening; lymphadenopathy at several sites was also reported, with histological correlation advised.

Esophagogastroduodenoscopy was performed under deep sedation with propofol target-controlled infusion using the Schnider pharmacokinetic model, titrated to a Modified Observer's Assessment of Alertness/Sedation (MOAA/S) score of 1–2. Supplemental oxygen was delivered through nasal cannulae incorporating capnographic sampling. A videolaryngoscope and flexible bronchoscope were prepared, with tracheotomy equipment immediately available. Figure 2 shows the oxygen interface, protective coverings, and vascular access. The postprocedural course was uneventful.

Table 1: Practical adaptations for endoscopy

Domain	Principal challenge	Practical response
Airway	Restricted access and mucosal trauma	Assess current anatomy; prepare a primary technique and rescue plan; limit attempts [7,8].
Ventilation	Obstruction or hypoventilation during sedation	Observe breathing and obtain exhaled carbon dioxide monitoring; verify unreliable sampling [9].
ECG	Trauma from adhesive electrodes	Use a conductive nonadhesive interface and confirm an interpretable signal [10].
Pulse oximetry	Scarred digits and exposed adhesive	Select an accessible site and isolate adhesive surfaces; verify the plethysmographic signal [10].
Blood pressure	Friction from the cuff	Use soft padding, correct cuff sizing, and scheduled measurements [9,10].
Vascular access	Difficult cannulation and fixation	Consider ultrasound; protect the catheter hub and use nonadhesive securement [10].
Positioning	Shear during transfers and movement	Lift rather than slide; pad contact points and support contractures [10].

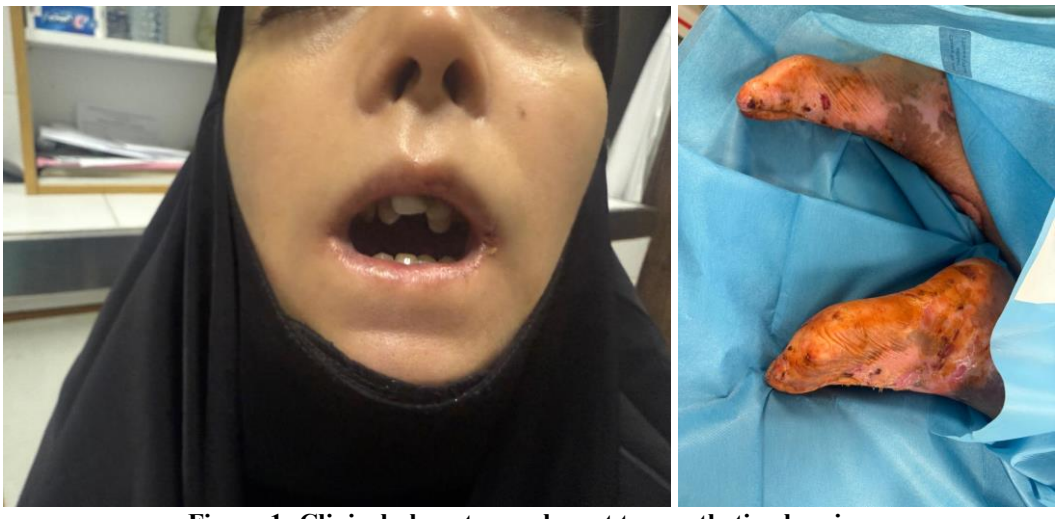


Figure 1: Clinical phenotype relevant to anesthetic planning

Left: oral opening and abnormal dentition. Right: cicatricial changes and marked deformity of the feet. These clinical features illustrate the need for individualized airway assessment and protection of fragile tissues.



Figure 2: Periprocedural setting

Periprocedural view showing protective coverings, the nasal oxygen interface, vascular access and suitable pulse oximeter.

DISCUSSION

Persistent dysphagia after repeated dilation creates both nutritional and procedural concerns. It should prompt reassessment of stricture anatomy and the objective of any further intervention. The 2025 EB esophageal guideline favors radiological characterization, discourages endoscopy solely to establish a stricture diagnosis, and conditionally favors fluoroscopically guided balloon dilation. Endoscopic assistance remains appropriate in selected circumstances according to anatomy, resources, and expertise. After unsuccessful treatment, the procedural objective and choice of approach require multidisciplinary reassessment [3].

Repeated dilation can be effective in expert centers. Anderson and colleagues reported 231 balloon dilations in 24 children and adolescents; adverse events attributable to dilation occurred after 10.0% of procedures, without procedure-related perforation, serious bleeding, or death. Most strictures were proximal, whereas the report in the present patient described distal narrowing. This difference reinforces the need for individualized anatomical assessment. The cohort cannot explain why the three procedures in this patient did not improve symptoms [4].

DEB can combine microstomia, reduced tongue mobility, scarring, abnormal dentition, and cervical contractures. Instrumentation may additionally provoke mucosal injury and bleeding. Adult airway anatomy may deteriorate over time, making previous uncomplicated anesthesia an incomplete predictor of current difficulty. Severe esophageal disease also raises aspiration concerns that are not eliminated by routine fasting [5].

Intrinsic laryngotracheal stenosis should not be assumed merely from the diagnosis of DEB. Registry data show that major tracheolaryngeal disease occurs particularly in junctional EB during childhood. Nevertheless, stridor, dysphonia, or unexplained respiratory symptoms warrant targeted evaluation. Distinguishing anatomical intubation difficulty from intrinsic airway obstruction helps avoid transferring subtype-specific risks indiscriminately to the present patient [6].

The choice between spontaneous breathing with sedation and a secured tracheal airway should account for aspiration risk, anticipated intervention, rescue access, and expertise. Preserving spontaneous ventilation does not guarantee airway patency. For an anticipated difficult intubation accompanied by aspiration risk, difficult ventilation, intolerance of apnea, or difficult emergency invasive rescue, the ASA difficult-airway guideline supports consideration of awake intubation. Oxygenation, limited attempts, an explicit failed-airway plan, and a planned extubation strategy are central safeguards [7]. In the present patient, TCI propofol sedation was accompanied by capnography and immediate availability of advanced airway equipment. This represents the approach used in this individual case rather than a general recommendation for patients with severe esophageal disease.

Flexible bronchoscopic intubation is particularly useful when oral access is severely restricted. Video laryngoscopy is an option only when mouth opening and tissue tolerance permit insertion. A 2026 pediatric report describes successful video laryngoscopic intubation for esophageal dilation,

demonstrating feasibility in a selected patient rather than superiority over bronchoscopy. Neither that experience nor a nasal route can be generalized to all adults; device choice must reflect the individual anatomy and operator experience [8].

A MOAA/S score of 1–2 is consistent with deep sedation and requires preparedness for airway obstruction, apnea, and progression to general anesthesia. Availability of advanced airway equipment complements a rehearsed rescue plan and skilled personnel [7,12]. A TCI target is a model-predicted concentration rather than a measured blood concentration; model, target compartment, and units must be specified before a numerical setting can be interpreted or reproduced.

Skin fragility changes how monitoring is applied. It does not remove the need to detect hypoxemia, hypoventilation, arrhythmia, or hypotension. Current ASA standards specify continuous ECG display and blood pressure and heart-rate assessment at least every five minutes during anesthesia. They also emphasize exhaled carbon dioxide surveillance, including during moderate or deep sedation, and quantitative carbon dioxide monitoring after placement of a tracheal or supraglottic airway. Exceptional departures require explicit clinical justification and documentation [9].

Practical adaptations include nonadhesive conductive interfaces for ECG electrodes, a pulse-oximeter sensor with no exposed adhesive, and a correctly fitted blood-pressure cuff over soft padding. Vascular access should be secured without standard adhesive contact; ultrasound may reduce repeated punctures. Transfers should minimize sliding, and equipment should not rest directly on fragile skin. These measures are summarized in Table 1 [10]. These are literature-based adaptations, not a record of measures confirmed in this patient.

Respiratory surveillance deserves particular attention during endoscopy, where the instrument and oxygen delivery system can impair capnographic sampling. A poor waveform requires assessment of sampling and clinical ventilation. Supplemental oxygen and an acceptable saturation value do not establish adequate alveolar ventilation; respiratory observation and carbon dioxide monitoring provide complementary information [9].

Brooks Peterson and colleagues described 202 general anesthetics in 37 patients managed through a specialist pathway; 75.7% had DEB. New skin or mucosal injury occurred in 4% of encounters and desaturation in 21.3%, with no major adverse events. The substantial frequency of desaturation is clinically relevant even when major complications are uncommon. This retrospective, predominantly pediatric experience supports careful preparation but cannot establish comparative safety between anesthetic techniques [11].

Some published series and case reports omitted ECG or other monitoring because of skin disease [8,11]. Those practices describe specific institutional decisions; they should not be converted into a general recommendation to omit surveillance when nonadhesive alternatives are available [9,10].

Recovery should be planned as carefully as induction. If intubation is used, emergence should minimize coughing and agitation while accounting for aspiration and difficult reintubation. Postanesthetic assessment should address new oral or cutaneous injury, respiratory symptoms, pain, and nausea [5]. In the present patient, persistent dysphagia also makes the subsequent feeding plan clinically important.

The educational value of this case lies in the convergence of risks during a commonly performed procedure. It illustrates why the visible skin phenotype should prompt a broader assessment of mucosal disease, airway access, and monitoring feasibility. The supporting literature remains largely observational, with variable subtypes, ages, procedures, and definitions of adverse events. Recommendations should therefore be interpreted as a framework for individualized care rather than proof of one optimal anesthetic technique.

CONCLUSION

Dystrophic epidermolysis bullosa can transform upper gastrointestinal endoscopy into a complex anesthetic encounter. In this 20-year-old woman with persistent dysphagia after three unsuccessful dilation procedures, TCI propofol sedation with nasal oxygen, capnography, and advance airway-rescue preparation was followed by an uneventful recovery. The broader lesson is the need to integrate airway and aspiration assessment, reliable monitoring, and protection from mechanical tissue injury. Technique selection remains individual and should reflect procedural requirements, airway anatomy, and the ability to provide prompt rescue.

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