orphanaesthesia

Anesthesia recommendations for

Epidermolysis bullosa

Disease name: Epidermolysis bullosa

ICD 10: Q81.0, Q81.2, Q81.8

ORPHAcode: 303, 304, 305, 2908

Synonyms: -

Disease summary: Epidermolysis bullosa (EB) is a heterogeneous group of inherited rare skin diseases characterized by extremely fragile skin and minimal shear force-induced blister and ulcer formation of the skin and mucosa. These diseases are caused by mutations in the genes for structural proteins of the skin basement membrane zone such as keratins, type VII collagen, or laminin 332. At least 16 different gene mutations result in more than 30 clinical subtypes of EB with a wide spectrum of clinical presentations. The incidence including all types is estimated at 19 per 1 million births.

Milder forms present with localized blistering mostly on the hands and feet after minor trauma, healing without scarring. In severe disease, generalized blistering with consecutive multisystemic involvement occurs at birth or shortly after. Blisters can form within the oral cavity, on the external surface of the eye, within the mucosa of the respiratory, gastrointestinal or genitourinary tract. Besides painful blistering, patients often present with secondary problems like pruritus, scarring and contractures. Severe microstomia, ankyloglossia, esophageal stenosis as well as significant dental decay may occur. Anemia, recurrent infections, cardiomyopathy, malnutrition, amyloidosis, renal failure and squamous cell carcinoma are concomitant morbidities reducing life expectancy [1,2,3,4,5,9,26].

Depending on the skin level of the blister formation, there are four major types of EB, all of which include multiple subtypes:

1) Epidermolysis bullosa simplex (EBS)

Blistering occurs within the epidermis. Lack of adhesion of the skin directly above the basement layer. The most common proteins affected are keratin 5 and 14, which are found within the basal keratinocytes [5]. Localized or generalized blistering may develop, usually no scarring. Generally milder than other types of EB.

A subtype of EBS caused by mutations of the kelch-like protein 24 (KLHL24) within the basal keratinocytes has recently been associated with dilated cardiomyopathy, which can be present without skin lesions [5].

This is the most common type of EB, accounting for 70% of cases. EBS usually is inherited in an autosomal dominant manner.

2) Junctional epidermolysis bullosa (JEB)

Blisters develop within the lamina lucida (the mid-portion of the skin basement membrane zone). Localized or generalized forms may develop, in many cases involvement of the mucosa resulting in failure to thrive, gastroesophageal reflux disease, constipation, protein-losing enteropathy. Patients often present with laryngeal and tracheal lesions leading to glottic and tracheal stenosis [7], ocular manifestations (lagophtalmos, ectropion, corneal abrasion, scarring, vision loss), poor dentition. Blisters tend to heal with scarring. The cumulative risk of dilated cardiomyopathy is 1.15%. Patients with severe JEB have a highly reduced life expectancy, most do not survive childhood [1,6].

This is the most severe type of EB, accounting for about 5% of cases, inherited in an autosomal recessive manner.

3) Dystrophic epidermolysis bullosa (DEB)

Autosomal dominant or recessive mutations in the gene encoding type VII collagen. These defects lead to blister formation beyond the basal membrane zone, always healing with scarring. Joint contractures, fusion of the fingers and toes (pseudosyndactyly), severe oral mucous membrane involvement and narrowing of the esophagus, gastroesophageal reflux disease, constipation, ophthalmic manifestations (lagophtalmos, ectropion, corneal abrasion, vision loss), poor dentition are common in severe types of DEB. Tracheolaryngeal stenosis may be found in 5% of patients in their mid-twenties. Dilated cardiomyopathy may develop in about 4.5% of patients with diagnosis at the mean age of 12 years [1].

Milder and more severe types of the disease are described. Onset usually is at birth, but a later onset in milder forms can be observed as well.

4) Kindler syndrome

Autosomal recessive inherited gene mutations encoding KIND1, a component of adhesion contacts in basal keratinocytes. Blistering may occur at multiple levels within the basement membrane zone or in skin layers beneath it. Generalized blistering is present at birth, development of characteristic poikilodermatous pigmentation and photosensitivity later, healing with atrophic scarring. Rare subtype of EB [4].

Therapy (all types): Strict prevention of friction and trauma is essential to avoid new blister formation, as healing is difficult. Gentle pressure on the skin is less problematic. Wounds, infections and strictures are managed symptomatically. Causal therapy is not yet available. Local cell-based gene therapy (beremagene geperpavec) for application on blisters, FDA approved for epidermolysis bullosa dystrophica since 2023, facilitates wound healing. However, high costs limit general accessibility. IL-4/13 inhibitors and Janus kinase inhibitors show promise for treating intense pruritus. Further research on modulating gene therapy and immunotherapy is going on [8].

Diagnosis may be incorrect; if uncertainty exists, the diagnosis should be re-evaluated.

Every patient is unique; individual circumstances must always guide clinical care.

Medicine is in progress; new clinical knowledge may not be yet reflected in this recommendation.

Recommendations are not rules or laws; they provide a framework to support clinical decision-making. Although this recommendation has passed a structured review process, it does not meet the formal criteria of a guideline.

Translations may not always reflect the most recent updates of the English version.

Find more information on the disease, its centers of reference and patient organizations on Orphanet: www.orpha.net

A	**AIRWAY / ANESTHETIC TECHNIQUE**	Avoid shear forces to skin and mucosa, as new blisters can occur. Gentle pressure on the skin is less problematic. Lubrication of all airway devices such as face mask, laryngoscope blade, endotracheal tube (ETT), laryngeal mask airway (LMA). Silicone-based dressings around the mouth opening are useful to avoid new blister formation in case of mask ventilation. Choose ETT or LMA one size smaller than predicted by standard formulas. Scarring can lead to microstomia, ankyloglossia, obstruction of the nostrils, joint contractures (neck, jaw), poor dentition. Blister formation and scarring (i.e., stenosis) at the level of the larynx or trachea are rare but occur in cases of EB junctionalis and severe cases of EB dystrophica. Equipment for difficult airway management should be available. Use apneic oxygenation for fiberoptic intubation. Asleep fiberoptic endotracheal intubation mostly remains the preferred technique for securing the airway if requested. Consider aspiration risk, if esophageal strictures are present. Consider anesthesia techniques maintaining spontaneous ventilation without airway instrumentation in order to avoid trauma to the airway structures.
B	**BLOOD PRODUCTS (COAGULATION)**	Consider hemoglobin value, blood coagulation profile and provision of blood products, if large areas of chronic wounds are present. Risk of anemia, chronic infections, malnutrition, renal impairment.
C	**CIRCULATION**	Consider cardiomyopathy, especially in dystrophic EB and EB simplex (recent echocardiography). Consider hypovolemia, if malnutrition or chronic infections are present. Consider difficult venous access.

D	**DRUGS**	Be aware of home medication for chronic pain and itching. Consider oral premedication for patient compliance and prevention of involuntary movements. Nasal premedication is possible, if nasal mucosa is not affected. Topical anesthesia can be applied without an adhesive dressing for venous cannulation. All general (GA) and regional anesthesia (RA) techniques can be applied according to their specific indications and contraindications. Aim for smooth induction of anesthesia and emergence to avoid coughing and uncontrolled movements.
E	**EQUIPMENT**	Silicone-based foam on the skin instead of any adhesive dressing is mandatory. Prefer clip-on pulse oximeters, no self-adhesive probes. Cut off the adhesive part of ECG electrodes and fix them with silicone-based mesh. Use cotton wool padding underneath the blood pressure cuff or tourniquet. Use lubricants for all airway devices. Use eye lubricants for cornea protection. Consider transillumination or ultrasound for venous access. Any shear forces must be avoided in transportation. Careful padding on the operation table. Preparing a dedicated trolley with the necessary equipment is advisable. Parents are very helpful to move the child: trolley to operating table etc.

Typical surgery and procedures

- Change of dressings, skin biopsy
- Dental surgery (dental extraction and conservation)
- Ophthalmic surgery
- Plastic surgery (repair of pseudosyndactyly, surgery to contractures, excision of squamous cell carcinoma, skin grafting)
- General surgery, esophagoscopy and -dilatation, gastrostomy, fundoplication, circumcision

Type of anesthesia

The subtype and the severity of disease and comorbidities require an individual approach to anesthesia management. Patients with severe disease presenting for elective procedures should be managed in a specialist center with multidisciplinary experience concerning EB.

Institutional protocols should be developed to care for these patients (wound dressings, pain management, airway management etc.) [3,9,10].

Meticulous planning of each anesthetic procedure is essential to prevent formation of new blisters.

If possible, prefer deep sedation with analgesia and/or regional anesthesia (RA) techniques due to the possibility of severe airway complications in case of new blister formation or due to pre-existing difficult airway due to scarring. Maintaining spontaneous ventilation without touching the airway is a preferable option for short interventions.

All types of general anesthesia (GA) (balanced techniques / total intravenous anesthesia, ketamine) have been described [11,12,13,14].

Peripheral RA and neuraxial anesthesia techniques can be performed safely, if there is no infection at the puncture site. Ultrasound guidance is very helpful. However, local skin infiltration should be avoided.

Topical local anesthesia (EMLA®, Ametop) can be applied on unaffected skin, as long as no adhesive dressing is used [12,15].

Necessary additional preoperative testing (beside standard care)

- History taking (records of previous anesthesia, presence of gastroesophageal reflux, muscular dystrophy, difficult airway, steroid therapy, pain therapy, renal dysfunction, infections)
- Blood values (blood cell count, renal parameters)
- Echocardiogram and electrocardiogram (if cardiomyopathy is assumed)
- Consultation of specialists: (pediatric) dermatologist, wound care specialist, pain service, ophthalmologist, dentist, nutritionist, physiotherapist, occupational therapist

Particular preparation for airway management

Recurrent blister formation and scarring can lead to microstomia, poor mouth opening, immobile tongue and esophageal webs, obstruction of the nostrils, nasopharyngeal fibrosis, tracheal stenosis, neck contractures. There is a considerable risk of blister formation at the level of the oropharyngeal airway in case of airway management. Airway difficulties may increase over time [16]. Laryngeal and tracheal involvement in EB occurs in 5% of patients with DEB in their mid-twenties, in up to 40% in patients with severe JEB at the age of 6 years. [1].

Thorough lubrication of face mask, laryngeal mask airway (LMA), endotracheal tube (ETT), laryngoscope is essential [2,3,12,15,17].

In severely affected patients, preventive coverage of certain areas of the face (e.g., nose, cheeks, chin) with special non-adhesive dressings (e.g., Mepilex® transfer) is recommended [12,17].

The ETT should be chosen half to one size smaller than predicted by standard formulas, provided with a softer cuff [9].

Equipment for predicted difficult airway should be available: e.g., video-laryngoscope, fiberoptic bronchoscope. Asleep nasal fiberoptic endotracheal intubation under passive nasal oxygen insufflation seems to be the preferred technique for securing the airway in severe cases, as the nasal mucosa is composed of respiratory epithelium, which is more resistant to shearing forces compared with the oral mucosa [9,18]. Transnasal humidified rapid-insufflation ventilatory exchange (THRIVE) can provide extended oxygenation without the need of bag-mask ventilation [19].

Many patients with less severe EB, however, can be intubated safely orally via direct laryngoscopy or video-laryngoscopy [2,3]. Mask ventilation is usually not difficult. Oropharyngeal tubes should be avoided.

The ETT should be fixed using a non-adhesive technique.

An LMA should be used one size smaller than predicted by standard formulas and intracuff pressure should be monitored (< 40 cmH2O). Well lubricated LMAs have been used safely but may initiate new intraoral blisters especially in case of microstomia [9,12,22]. The use of an LMA may be appropriate in airway emergencies. However, LMAs should generally be used restrictively because of the larger area of contact with the mucosal surface and the risk of shear forces during placement or removal [9].

With appropriate precautions, safe endoscopic laryngeal surgery in patients with JEB has been reported in a case series [23,24].

Particular preparation for transfusion or administration of blood products

Some severe forms of epidermolysis bullosa can result in a transfusion-dependent anemia. Causes include blood and iron loss from wounds, chronic inflammation, malnutrition and problems absorbing iron due to the effects EB has on the gastrointestinal tract. Oral and intravenous iron can be given, often paired with erythropoietin shots. If necessary, a blood transfusion is another option. Common risks and contraindications of each blood transfusion have to be considered [20].

Particular preparation for anticoagulation

Not reported.

Particular precautions for positioning, transportation and mobilization

The most important task for transport or mobilization of the patient is to maintain the integrity of the skin, avoiding friction, secondary pressure and trauma. Thus use a “lift and place” approach during transfers. If possible, the patient should move independently between surfaces [6,10,21]. The help of parents is very helpful.

The operating table needs to be extensively padded, and the patient has to be transferred very carefully. Any pulling of the patient over surfaces during transfer in and out of the operating room has to be avoided.

The operating room needs to be warmed in order to prevent heat loss of the patient due to the extent of skin lesions and malnutrition.

Interactions of chronic disease and anesthesia medications

General medication should be taken as usual. Be aware that many patients with EB take an opioid medication for chronic pain. Attention should be paid to infection prophylaxis.

Chronic local steroid therapy is often used to reduce inflammation: beware of subclinical steroid insufficiency that could become apparent during the perioperative period (hypotension, hypoglycemia).

Anesthetic procedure

Agitation and uncontrolled movement during induction can lead to new skin damage.

A preoperative sedative medication could be appropriate.

As the intravenous access can be difficult, consider the use of ultrasound or transillumination technique for establishment of intravenous access.

A rapid and non-traumatic intravenous induction is advantageous, but an inhalational induction is also possible. Mask ventilation usually is not difficult, as the tongue often is tethered and does not tend to fall back, but prolonged facial manipulation can lead to new blisters. The skin area should be protected by a silicone-based foam dressing [9,12,14].

To avoid new blisters, use Vaseline or any lubricant for face masks, laryngoscope, ETTs, LMAs and stomach tube.

Oropharyngeal suctioning should be done carefully without contact with the mucosa.

If the patient has gastroesophageal reflux, a rapid sequence induction and intubation is indicated. Due to a smaller mouth opening, ankyloglossia (little tongue movement), adhered fixed epiglottis, less reclination and possible tracheal stenosis, be prepared for a difficult airway as described above.

For eye protection, use a moisturizing ophthalmic gel, preferably free of preservatives or lanolin. After application of the gel, cover the eyes with moistened gauze to protect them from mechanical trauma. Take care that patients will not wake up with blurred vision and rub their eyes after extubation. Risk of corneal abrasion [14,15].

Total intravenous anesthesia may be useful in reducing agitation and emesis in the recovery room. Non-depolarizing muscle relaxants sometimes show prolonged duration of action due to hypoalbuminemia and low muscle mass [15]. Succinylcholine has been used successfully [11,12,22]. Avoid fluid and heat loss and consider a sophisticated pain treatment.

Although GA is mostly used, RA is advisable as a basic principle, often in combination with sedation, as airway management can be avoided. Single shot and continuous peripheral nerve blocks as well as central neuraxial blocks have been performed successfully without additional risk. Subcutaneous infiltration with local anesthetics should be avoided, as new blisters can

occur. For all these procedures, rubbing or wiping the skin for disinfection should be avoided, whereas patting the skin with a moist wipe is usually well tolerated [21,25].

Smooth awake extubation without coughing and uncontrolled movements is advisable [9].

Particular or additional monitoring

Standard monitoring is sufficient, adapted to the surgical intervention. Any adhesive is contraindicated because it may cause new blisters. Generally, consider minimizing monitoring whenever possible to avoid further harm to the patient, but use it, if medically indicated [2,3,4,9,17].

The adhesive part of ECG leads should be cut off and the electrodes fixed by silicone-based tape.

Clip-on pulse oximeters can be used safely.

The self-adhesive part of pediatric pulse oximeter probes must be removed, the probe itself can be fixed by silicone-based tape. Alternatively, the adhesive part may be covered by the adhesive site of a Tegaderm® dressing. The non-adhesive site of the Tegaderm® thereafter may be secured on the skin by silicone-based tape (Mepilex®) [17,21].

Intravenous catheter and any other device should be fixed with non-adhesive techniques like silicone-based products (Mepilex® transfer, Mepitac®).

A layer of cotton-wool padding should underlay the blood pressure cuff or the tourniquet.

Arterial lines should be sutured in place [21]. More liberal indication for placement of arterial lines under US guidance in order to protect the skin from non-invasive blood pressure measurement.

Avoid invasive temperature probes if possible. A lubricated axillary probe may be preferred.

Possible complications

- New blisters, especially oropharyngeal and periglottic lesions
- Difficulty obtaining venous access due to contractions and multiple scars
- Difficult airway due to small mouth opening, laryngeal or tracheal stenosis
- Dysphagia, esophageal stenosis and reflux
- Corneal lesions
- Anemia, hypovolemia
- Septic complications
- Urinary retention after removal of urinary catheter
- Difficult pain therapy

Postoperative care

Excellent analgesia is important to prevent excessive movements and new skin trauma. A multimodal approach using nonsteroidal analgesics and opioids by the intravenous route is the most convenient method. High doses of analgesics may be required. Continuous RA techniques are advisable, if indicated. Use of rectal suppositories is discussed controversially, as the rectal manipulation may provoke blisters [27].

Many patients suffer from severe chronic itch. Therapeutic options are antihistamines, gabapentin, pregabalin, serotonin-noradrenaline reuptake inhibitors and behavioral interventions. Recently available medications like IL-4/13 inhibitors and Janus kinase inhibitors show promise for treating intense pruritus [8].

In case of emergence delirium, immediate sedation is recommended, as uncontrolled movements easily cause new blisters.

Swallowing of oral medication/nourishing can be painful after airway manipulation and due to pre-existing significant esophageal stenosis. Many patients are not able to swallow pills or capsules.

Oxygen masks with sharp edges should be strictly avoided. Prolonged surveillance in the recovery room should be planned, if airway management was performed, as new intraoral blisters may occur.

Disease-related acute problems and effect on anesthesia and recovery

- Systemic inflammatory reactions
- Septic complications
- Acute esophageal obstruction, dysphagia
- Acute upper airway obstruction due to formation of new blisters in the upper airway tract [6]

Ambulatory anesthesia

Small procedures can be realized in an experienced ambulatory setting. If airway manipulations are considered, an ambulatory management is not recommended because of the risk of formation of new intraoral blisters. Generally, pain management can be challenging.

Obstetrical anesthesia

Antenatal anesthetic assessment is recommended for patients with moderate or severe EB [28].

Pregnancy itself is not affected by the disease. The severity of skin fragility, however, may change to the better as well as to the worse during pregnancy. Anemia is found in about 30% of pregnancies, especially in patients with more severe EB [29]. Vaginal delivery is the preferred mode and possible for most patients. Labor complication rates in patients with mild

EB are not increased [29,30]. Perineal tears rate increases to about 15% with severity of EB due to fragility of the skin [29,30].

Blisters of the vaginal mucosa have been described after delivery, but vaginal delivery does not appear to increase the risk of subsequent vaginal scarring or stenosis even in patients with severe EB. Both episiotomies and perineal lacerations tend to heal well. The cesarean section rate is increased to about 50% in patients with severe EB, mostly due to preexisting vulvovaginal involvement [28].

Many case reports describe successful cesarean delivery without negative consequences for mother and child. RA (spinal/epidural) remains the preferred technique and can be performed safely, if there are no infected blisters at the puncture site [28,29,31]. If urinary catheter is necessary, it needs to be well lubricated before insertion, and a smaller size than usual should be chosen [21]. Some patients with EB have strictures of the urinary tract, which might render a catheter insertion impossible.

If difficult airway management is anticipated, early insertion of an epidural catheter may be considered to prevent GA in case of emergency [5].

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Network information

EB international patient advocacy and support network: www.debra-international.org

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