

# orphan**a**nesthesia

## Anaesthesia recommendations for patients suffering from **Epidermolysis bullosa**

**Disease name:** Epidermolysis bullosa

**ICD 10:** Q81

**Synonyms:** -

Epidermolysis bullosa (EB) is a heterogeneous group of inherited rare diseases, which are characterized by trauma-induced blister formation of the skin and mucosa. The underlying cause is a functional deficiency of structural proteins of the dermo-epidermal junction. Depending on the level of the blister formation, EB is divided into EB simplex (intra-epidermal), junctional EB (within the lamina lucida), dystrophic EB (below the lamina lucida) and Kindler syndrome (variable level of split formation). Besides different distinct blister formation and pain symptoms secondary problems like anaemia, oesophageal stenosis, cardiomyopathy or squamous cell carcinoma may occur. Furthermore, many patients present with severe microstomia and ankyloglosson as well as significant dental decay.

Since causal therapies are not available, strict prevention of friction and trauma is essential to avoid blister formation.

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Medicine in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

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### Typical surgery

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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.

### Type of anaesthesia

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- General anaesthesia (balanced anaesthesia, total intravenous anaesthesia)
- Analgosedation
- Spinal anaesthesia
- Epidural anaesthesia
- Combined spinal and epidural anaesthesia
- Peripheral nerve blocks
- Local application of EMLA®.

### Necessary additional diagnostic procedures (preoperative)

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History taking (records of previous anaesthesia, presence of gastro-oesophageal reflux, muscular dystrophy, difficult airway, steroid therapy, renal dysfunction)

Blood values (count, renal)

Echocardiogram and electrocardiogram (if cardiomyopathy is assumed)

Consultation of specialists: (paediatric) dermatologist, wound care specialist, pain service, ophthalmologist, dentist, nutritionist, physiotherapist, occupational therapist.

### Particular preparation for airway management

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Evaluation of a difficult airway (history taking, records of previous anaesthesia)

Thorough lubrication of face mask, laryngeal mask airway (LMA), endotracheal tube (ETT), laryngoscope

In very affected patients coverage of certain areas of the face (e.g. cheeks, chin) with special non-adhesive dressings (e.g. Mepilex transfer) is recommended

LMA one size smaller than predicted by standard formulas

Cuffed endotracheal tube (ETT) half to one size smaller than predicted by standard formulas

Equipment for predicted difficult airway: E.g. video laryngoscope, fiberoptic bronchoscope.

### **Particular preparation for transfusion or administration of blood products**

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Some severe forms of epidermolysis bullosa can result in a transfusion dependent anaemia. The cause is unknown, but many factors take part in it like blood and iron loss from wounds, chronic infection, 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 erythropoetin shots. If necessary a blood transfusion is another option. Common risks and contraindications of each blood transfusion have to be considered.

### **Particular preparation for anticoagulation**

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Not reported.

### **Particular precautions for positioning, transport or mobilisation**

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The most important task for transport or mobilisation of the patient is to maintain the integrity of the skin, avoiding friction, secondary pressure and traumata. 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 an out of the operating room has to be avoided.

### **Probable interaction between anaesthetic agents and patient's long term medication**

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Not reported.

General medication should be taken as usual. Take care that many patients with EB take an opioid medication for chronic pain. Attention should be paid on infection prophylaxis.

### **Anaesthesiologic procedure**

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Agitation and uncontrolled movement during induction can lead to new skin damage. For this reason a good preoperative sedation is essential. A rapid and nontraumatic intravenous induction is advantageous, but an inhalational induction is also possible. As intravenous access often is difficult consider the use of ultrasound technique for establishment of intravenous access. To avoid new blisters use vaseline or any lubricant for face masks, laryngoscope, endotracheal tubes and stomach tube. Endotracheal tube should be fixed with nonadhesive technique. If the patient has reflux, a rapid sequence induction is indicated. Due to a smaller mouth opening, ankyloglossia (little tongue movement), adhered epiglottis, less reclination and possible tracheal stenosis be prepared to a difficult airway. The endotracheal tube should be taken half number smaller than anticipated to age and be minimally blocked. Well lubricated laryngeal mask can be used but may initiate new blisters. Nasal fiberoptic intubation should be avoided, the oral access should be chosen. Oropharyngeal tubes should be avoided. Total intravenous anaesthesia may be useful in possibly reducing agitation and emesis in the recovery room. Succinylcholine has been used successfully. Non depolarising muscle relaxants sometimes show prolonged duration of action due to hypoalbuminemia and low muscle mass. One needs to be very gentle and careful when

suctioning the stomach and oropharynx before extubating as this easily causes new wounds. Avoid fluid and heat loss and consider a sophisticated pain treatment. Although general anaesthesia is mostly done, regional anaesthesia is also possible. Single shot and continuous nerve blocks as well as central neuroaxial blocks have been performed successfully without additional risk. Subcutaneous infiltration with local anaesthetics should be minimised 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. If general anaesthesia can be avoided, one should prefer analgesedation or regional anesthesia techniques due to the possible airway complications.

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#### Particular or additional monitoring

Standard monitoring is sufficient, adapted to the surgical intervention. Any adhesive is contraindicated because it may cause new blisters. ECG, intravenous catheter and any other device should be fixed with nonadhesive technique like silicon 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. For eye protection use a moisturizing ophthalmologic gel, preferably free of preservatives or lanolin. After application of the gel cover the eyes with moistened gauze to protect them from mechanic trauma. Take care that the patient will not wake up with blurred vision and rub at his eyes after extubation. Risk of corneal abrasion. Generally consider to minimize monitoring whenever possible to avoid further harm to the patient.

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#### Possible complications

- New blisters, especially oropharyngeal and periglottic
- Difficulty getting venous access due to contracted fingers and multiple scars
- Difficult airway due to small mouth opening or tracheal stenosis
- Dysphagia, esophageal stenosis and reflux
- Anaemia
- Difficult pain therapy.

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#### 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 analgetics may be required. Regional anaesthesia should be considered whenever possible. Rectal suppositories are not recommended in first line because of the risk of rectal wounds, but successful use has been reported. If used, they should be well lubricated with a jelly. Swallowing of oral medication / nourishing can be painful after airway manipulation and due to often significant oesophageal stenosis, many patients are not able to swallow pills or capsules. Oxygen masks with sharp edges should be strictly avoided. In case of emergence delirium aggressive sedation is recommended.

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### Information about emergency-like situations /Differential diagnostics

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*caused by the illness to give a tool to distinguish between a side effect of the anaesthetic procedure and a manifestation of the disease*

- Systemic inflammatory reactions
- Septic complications.

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### Ambulatory anaesthesia

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

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### Obstetrical anaesthesia

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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. Vaginal delivery is the preferred mode and possible for most patients, but blisters of the vaginal mucosa have been described after delivery. Regional anaesthesia (spinal/epidural anaesthesia) can be performed safely, if there are no infected blisters at the puncture site. If urinary catheters are necessary, they need to be well lubricated before insertion. Some patients with EB have strictures of the urinary tract which might render a catheter insertion impossible.

Due to frequently difficult airway management, an early insertion of an epidural catheter should be considered so that general anaesthesia in case of emergency can be avoided.

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