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Bullous pemphigoid

Congenital hypothyroidism

orphan**anesthesia**

a project of the German Society
of Anaesthesiology and Intensive Care Medicine

SUPPLEMENT NR. 7 | 2020

OrphanAnesthesia –

ein krankheitsübergreifendes Projekt des Wissenschaftlichen Arbeitskreises Kinderanästhesie der Deutschen Gesellschaft für Anästhesiologie und Intensivmedizin e.V.

Ziel des Projektes ist die Veröffentlichung von Handlungsempfehlungen zur anästhesiologischen Betreuung von Patienten mit seltenen Erkrankungen. Damit will OrphanAnesthesia einen wichtigen Beitrag zur Erhöhung der Patientensicherheit leisten.

Patienten mit seltenen Erkrankungen benötigen für verschiedene diagnostische oder therapeutische Prozeduren eine anästhesiologische Betreuung, die mit einem erhöhten Risiko für anästhesieassoziierte Komplikationen einhergehen. Weil diese Erkrankungen selten auftreten, können Anästhesisten damit keine Erfahrungen gesammelt haben, so dass für die Planung der Narkose die Einholung weiterer Information unerlässlich ist. Durch vorhandene spezifische Informationen kann die Inzidenz von mit der Narkose assoziierten Komplikationen gesenkt werden. Zur Verfügung stehendes Wissen schafft Sicherheit im Prozess der Patientenversorgung.

Die Handlungsempfehlungen von OrphanAnesthesia sind standardisiert und durchlaufen nach ihrer Erstellung einen Peer-Review-Prozess, an dem ein Anästhesist sowie ein weiterer Krankheitsexperte (z.B. Pädiater oder Neurologe) beteiligt sind. Das Projekt ist international ausgerichtet, so dass die Handlungsempfehlungen grundsätzlich in englischer Sprache veröffentlicht werden.

Ab Heft 5/2014 werden im monatlichen Rhythmus je zwei Handlungsempfehlungen als Supplement der A&I unter www.ai-online.info veröffentlicht. Als Bestandteil der A&I sind die Handlungsempfehlungen damit auch zitierfähig. Sonderdrucke können gegen Entgelt bestellt werden.

OrphanAnesthesia –

a project of the Scientific Working Group of Paediatric Anaesthesia of the German Society of Anaesthesiology and Intensive Care Medicine

The target of OrphanAnesthesia is the publication of anaesthesia recommendations for patients suffering from rare diseases in order to improve patients' safety. When it comes to the management of patients with rare diseases, there are only sparse evidence-based facts and even far less knowledge in the anaesthetic outcome. OrphanAnesthesia would like to merge this knowledge based on scientific publications and proven experience of specialists making it available for physicians worldwide free of charge.

All OrphanAnesthesia recommendations are standardized and need to pass a peer review process. They are being reviewed by at least one anaesthesiologist and another disease expert (e.g. paediatrician or neurologist) involved in the treatment of this group of patients.

The project OrphanAnesthesia is internationally oriented. Thus all recommendations will be published in English.

Starting with issue 5/2014, we'll publish the OrphanAnesthesia recommendations as a monthly supplement of A&I (Anästhesiologie & Intensivmedizin). Thus they can be accessed and downloaded via www.ai-online.info. As being part of the journal, the recommendations will be quotable. Reprints can be ordered for payment.

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orphananesthesia

Anaesthesia recommendations for Bullous pemphigoid

Disease name: Bullous pemphigoid (BP)

ICD 10: L12.0

Synonyms: -

Disease summary: Bullous pemphigoid is an acquired, chronic, blistering autoimmune sub-epidermal bullous disease in which autoantibodies are directed against the component of the basement membrane zone of the skin [1]. It is characterised by the formation of cutaneous bullae on the skin and mucous membrane. The pathogenesis involves migration of inflammatory cells into sub-epithelial tissues due to the activation of complement caused by antigen-antibody reaction. The incidence of BP is 6–7 cases per million per year in the Western world. It usually affects elderly people with more than 60 years of age and is rare in children although childhood BP has been described [2].

One of the variants of the disease is cicatricial pemphigoid, which commonly involves the mucous membranes of the oropharynx, conjunctiva, nasopharynx, larynx, oesophagus, genitalia and anus. Bullous eruptions are followed by scarring [3].

The clinical course of the disease is one of exacerbations and remissions. The diagnosis is based on clinical presentation, histopathologic analysis, direct and indirect immunofluorescence microscopy on perilesional skin (DIF), analysis of staining patterns, and/or the characterisation of circulating autoantibodies [4].

There have been several reports of its association with other autoimmune skin/bullous diseases like pemphigus, pemphigoid, epidermolysis bullosa acquisita, dermatitis herpetiformis (Dhring), linear immunoglobulin-A disease, and multiple autoimmune syndrome [5]. Recently, some case reports and a review on its association with Acquired Haemophilia A (AHA) have been published. The mechanism is unclear, however, it is postulated that there may be a possible relationship between the 2 autoimmune diseases. Seventeen cases of BP associated with AHA, ages ranging from 24 to 88 years, have been documented [6].

The association of BP with malignancy is a matter of debate with no consensus. Despite several published case reports and trials, a definite association is lacking. Ogawa et al. found a significantly higher incidence of malignancy in patients with BP [7]. Other Asian authors have also reported a higher incidence of malignancy in their patients [8]; however, other studies done in Caucasians have failed to prove any statistically significant association [4].

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Treatment is with corticosteroids and immunosuppressive drugs [9] and occasionally antibiotics if an associated bacterial infection develops. Early disease can be managed by topical steroids but advanced disease requires systemic doses. In cases where steroids are contraindicated, treatment is tetracycline alone or combined with nicotinate. Cytotoxic drugs form the second line of treatment (azathioprine and methotrexate). A high dose of intravenous immunoglobulins (IVG) is indicated in resistant cases [10].

Medicine is in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong



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

Typical surgery

The patient can present for various types of surgery, both elective and emergency. Surgery in these patients includes restoration of function where severe deformity is present, e.g. laryngeal and oesophageal strictures and ophthalmological conditions. A patient may present as an emergency, e.g. for laparotomy (due to gastro-duodenal ulceration), or for Caesarean section, management of acute airway obstruction secondary to cicatricial laryngeal lesions, orthopaedic procedures, or other routine elective procedures [11,12].

Once laryngeal stenosis occurs, repeated endoscopic laser excision of scar tissue may be needed to maintain an adequate airway [13].

Type of anaesthesia

There are no definite recommendations for either general or regional anaesthesia.

Ketamine-based general anaesthesia without tracheal intubation and patient breathing spontaneously has been used in several BP patients. It provides rapid induction, preservation of airway reflexes and minimal trauma to the face and upper airway.

Despite concern about airway trauma, general anaesthesia with tracheal intubation can be used if considered safer for the patient [14]. Tracheal intubation in a similar condition, epidermolysis bullosa dystrophica, has been described in 131 patients without intraoperative or post-operative airway obstruction [15].

Whenever general anaesthesia requiring a secure airway (intubation/laryngeal airway) is unavoidable, protective measures must be ensured, e.g. a tracheal tube should be secured with soft cloth bandage rather than with adhesive tape. In patients with documented oral lesions, inserting a tracheal tube is a safer technique compared to a laryngeal mask airway because of the risk of bleeding from intraoral lesions [16].

The use of regional anaesthesia in these patients is controversial, the reason being that blister formation is a possibility at the site of epidural catheter insertion. These blisters can also interfere with epidural catheter fixation on the skin. However, regional anaesthesia has been described for surgeries as it avoids general anaesthesia and airway manipulation [17,18]. The use of tape to secure the epidural catheter to a large area of skin should be avoided. Spinal anaesthesia has been described for Caesarean sections and avoids the need for skin infiltration at the site of insertion [19].

Neuraxial opioids should be avoided as they are associated with pruritis, especially in cases involving morphine [20].

Necessary additional pre-operative testing (beside standard care)

- Complete blood count is indicated if patients are on myelosuppressive drugs like corticosteroids and azathioprine.
- Measurement of serum electrolytes, especially potassium and sodium, is indicated because potassium may be lost through skin lesions leading to hypokalaemia. Sodium can be deranged due to fluid loss from blisters and dehydration.

- Lung function tests are indicated if a supraglottic airway obstruction is present [21] (spirometry should be done).
- Hepatic function tests are indicated if the patient is on immunosuppressive drugs.
- Serum protein measurements should be performed in patients with oral blisters who are likely to have malnutrition.

Particular preparation for airway management

Airway issues:

- Patients may present with extensive tracheobronchial involvement with cough and dyspnoea [22]
- Vesicular eruptions leading to scarring which may occlude the upper airway and patient may present with stridor
- Pre-existing bullae in the oropharynx may restrict mouth opening
- Difficult intubation due to narrowing of laryngeal inlet
- Risk of haemorrhage is present secondary to airway manipulation
- Further risk of inducing new bullae formation with laryngoscopy and tracheal intubation.

Airway management:

- Oral intubation is preferable to the nasal route
- 1% hydrocortisone cream and vaseline-soaked gauze should be applied on face to avoid direct face mask contact as it poses the risk of severe bullae formation [23]
- Oral airway should be avoided
- Use an endotracheal tube smaller than indicated size
- Tracheal tubes should be secured with a soft cloth bandage
- Suction catheters should be well lubricated and suction pressure should be kept low
- Tracheostomy under local anaesthesia is indicated if the upper airway is complicated by severe ulceration, adhesions and deformity of oral mucosa, epiglottis and web adhesion of the vocal cords [24].

Particular preparation for transfusion or administration of blood products

In BP, autoantibodies are targeted against hemidesmosomal components BP 180 [3] and BP 2304 [25]. Therapeutic plasmapheresis is an effective treatment and has definite therapeutic advantages in some bullous disorders including bullous pemphigoid [26].

There is no evidence that BP is modified by BT [25].

Particular preparation for anticoagulation

There is no evidence in literature regarding any particular preparation for anticoagulation in BP.

Particular precautions for positioning, transportation and mobilisation

To avoid friction and pressure-induced trauma, careful positioning should be done. It is preferable to let patients position themselves on the operating room table as it will reduce pressure- and friction-induced trauma. Sheets should be free of creases. Gauze pieces soaked with hydrocortisone cream and vaseline may be applied to pressure areas like heels and elbows [14].

Interactions of chronic disease and anaesthesia medications

BP onset has no direct interaction with anaesthetic drugs but may be associated with the following adjuvant drugs that are likely to be used during the perioperative period. These are furosemide, ibuprofen, ACE inhibitors, metronidazole, spironolactone and some antibiotics (ampicillin, cephalosporins) [27].

Neuraxial opioids should be avoided as they may cause pruritis and worsen the skin condition.

Anaesthetic procedure

Anaesthetic implications for airway management have already been listed. Additional management points are:

- If patients are on regular corticosteroid therapy, they need to have a pre-operative loading dose of corticosteroid.
- Taping should not be done to protect the eyes.
- Peripheral vascular/venous access may be difficult in some cases where central venous cannulation is indicated and it should be fixed with skin stitches.
- Local infiltration of the anaesthetic agent is contraindicated because of the risk of skin sloughing and bullae formation.

- In case of needle insertion for spinal anaesthesia, an area devoid of skin lesions should be preferred.
- In difficult intubations, muscle relaxants should be avoided. A mixture of oxygen (O₂) with a low-density gas like helium (if available) is recommended (O₂ 20% and Helium 80%) to decrease the turbulence in flow.
- A clear polyethylene head hood should be used in a spontaneously ventilating patient.
- Opiates, thiopental sodium and propofol can be used without any complication.

Particular or additional monitoring

Care should be taken to avoid pressure and friction during the placement of monitoring devices, ECG pads, intravenous and arterial lines. IV lines and arterial lines should be fixed with sutures and gauze bandages [26]. Taping should be avoided.

Blood pressure cuffs should be well padded. Some authors prefer direct measurement of blood pressure in preference to non-invasive blood pressure.

Avoid temperature probe insertion in nasal and oral cavities.

Possible complications

Rough handling may cause new bullae formation and bleeding from raw areas.

Bullae may form in the trachea due to mechanical contact of tracheal tube balloon [28].

Subcutaneous emphysema may develop in narrowed laryngeal orifices due to high-pressure jet ventilation [29].

In patients with BP, traumatised skin often develops lesions in the area where the disease had not previously been seen. This is known as the Koebner phenomenon [20,30].

Post-operative care

Avoid blind pharyngeal suctioning at the end of the procedure.

Extubation should be gentle and the patient should be observed for post-operative stridor, new bullae formation and any difficulty in breathing.

Disease-related acute problems and effect on anaesthesia and recovery

Emergency-like situations may happen if cicatricial pemphigoid involves the oral cavity, epiglottis and vocal cords. Patients may present with stridor and dyspnoea due to narrowing of the laryngeal inlet. Obstruction is more obvious during inspiration, as the walls of the glottis are drawn together due to negative intratracheal pressure. Tracheostomy may be needed in advance in some cases.

Ambulatory anaesthesia

Ambulatory anaesthesia should only be done in patients with BP if mucous membranes are not involved in the disease process.

Obstetrical anaesthesia

Pregnancy can precipitate and aggravate BP especially during the 1st and 2nd trimester. However, in the third trimester, the clinical picture can improve due to an increased endogenous corticosteroid production by the chorion. Sepsis and skin infection at the site of local anaesthesia injections is possible. A choice of an area devoid of skin lesion is recommended and considered safe for performing a lumbar puncture.

Case reports have been published which show that single-shot spinal anaesthesia is safe for Caesarean section, however, general anaesthesia is not contraindicated if the oral mucosa is not associated with the disease [19].

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