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Case report

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ANESTHESIA FOR A NEONATE WITH PYLORIC ATRESIA-JUNCTIONAL EPIDERMOLYSIS BULLOSA SYNDROME: A CASE REPORT

SUMMARY

The paper describes the anesthetic management of a neonate with pyloric atresia – junctional epidermolysis bullosa (PA-JEB) syndrome. Anesthesia for the neonate with PA-JEB syndrome can stand for a serious challenge even to the most experienced anesthesiologists. Therefore, a few basic principles will help to organize the necessary procedures.

Shearing forces applied to the skin will result in bulla formation, while compressive forces to the skin are tolerated. Adhesive tape, adhesive ECG electrodes, adhesive pulse oximeter probes should not be used under any circumstances. Padding should be profusely used. Face masks should be lubricated with some emollient or coated with several layers of Vaseline gauze. All the instruments places into the mouth (laryngoscope, oropharyngeal airways) must be thoroughly lubricated with water based lubricant – do not lubricate with Lidocaine jelly. Pharyngeal suctioning should be avoided. Tracheal lesions do not appear after intubation probably because the trachea is lined with columnar epithelium. To reduce the risk of new laryngeal bullae formation, a tracheal tube a half to one size smaller than predicted may be necessary.

With maximal skin and mucous membrane protection, anesthesia in children with PA-JEB syndrome may be conducted with a few sequelae.

Key words: **PA-JEB syndrome, neonate, anesthesia**

INTRODUCTION

There are over 20 types of epidermolysis bullosa (EB) described, with three major subtypes: dystrophic EB (DEB), EB simplex (EBS), and junctional EB (JEB), with each broad category of EB containing several subtypes. Each type of EB is distinguished on the basis of the skin level in which the characteristic blistering occurs (figure 1).

There are three major subtypes of JEB: Herlitz, non Herlitz and JEB with pyloric atresia (1). JEB with pyloric atresia (JEB-PA), also known as Carmi syndrome (2), is a rare form of JEB having a poor prognosis. A novel subtype has been discovered recently - EBS associated with PA (3).

Pyloric atresia (PA) is a primary manifestation rather than a scarring process secondary to JEB – more properly called "PA-JEB" (4). Not uncommonly, patients with the PA-JEB syndrome present with erosions and/or subepithelial cleavage in the respiratory, gastrointestinal, and urinary tracts. In addition, certain facultative features are unique to PA-JEB, i.e. obstruction of the ureterovesical junction and high incidence of a peculiar form of aplasia cutis congenita (5). This autosomal recessive inherited disease is usually fatal within the first few weeks or months of life even following surgical correction of the intestinal obstruction (6).

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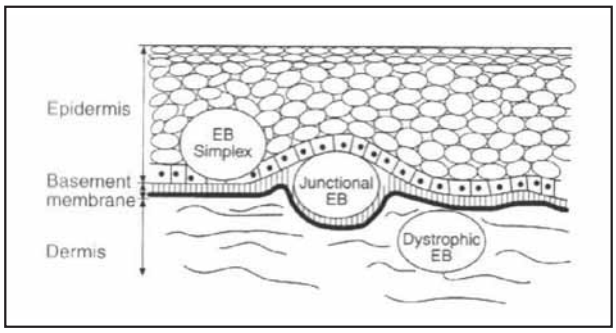

Figure 1. Level of cutaneous blisters in different types of epidermolysis bullosa

The main aspect of anesthetic management of these patients is a detailed knowledge of all the problems that anesthesiologist must face and solve to preserve the skin and mucosa integrity, avoiding the risk of severe complications (7).

CASE REPORT

A male neonate, born at term, with body weight 3.4 kg was admitted to our hospital on the 14th of June in 2003 (48 hours after the birth) for the evaluation of signs of Ileus meconialis susp. At the initial presentation baby was dyspneic, febrile with foamy secretion draining from the mouth. There were no signs of discharging of the bowels (stool). NG tube was placed, the initial clinical and laboratory evaluation performed (urea 13.9 mmol/L, glycemia 2.29 mmol/L, LDH 1008 U/L, electrolytes, total proteins, AST, ALT, gamma GT, amylase, blood gases parameters, as well as blood count were within the normal range). Radiologic examination (gastroduodenal passage) revealed no signs of the contrast in duodenum.

At the place where NG tube was fixated in the nose and underlying skin where intravenous cannula was secured, we noticed small blisters. We suspected that this was the case of the so-called "PA-JEB syndrome". Premedication (atropine) was i.m. administered - there are no reports of trauma after the i.m. route. In order to reduce the risk of skin damage, we moved the patient as little as possible. Anesthesia induction was inhalational (halothane) and we covered an anesthetic facemask with a paraffin gauze, as well the face under the chin. We lubricated the laryngoscope blade into the airway. The tracheal tube was 3.0 I.D. without cuff. Prior to extubation, pharyngeal suctioning was performed. We used pulse oximetry probe of the "clip-on" variety, and for the ECG monitoring we removed the adhesive part of the electrodes as much as we could. In order to measure the arterial pressure, we used sphygmomanometer (not-automated devices with potentials to generate high cuff pressure). The surgical procedure (gastro-duodenostomy latero-lateralis) and anesthesia course were uneventful. At the end of the surgery we noticed several bullae formation, one of which was on the right elbow (figure 2).

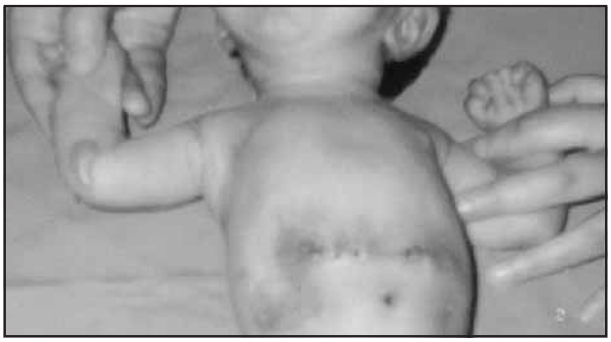

Figure 2. Blister on the right elbow noticed at the end of the surgery

DISCUSSION

Generally, management is one of the "no touch" principles (8). However, direct pressure to the skin is not as damaging as frictional or shearing forces. Therefore, adhesive dressing should never be used. Even positioning requires the outmost care to avoid moving of the sheets beneath the patient.

Minimal manual venous occlusion to aid peripheral vein cannulation should be performed by support staff. Intravenous cannulae should be secured with a non-adhesive dressing.

Even a minimal monitoring is not always possible, and these babies are often poorly monitored. To monitor the oxygen saturation, a probe of the 'clip-on' variety (adult clip-on probe) placed on a digit or the ear lobe is ideal. If only pulse oximetry probes available are self-adhesive, then the digit should be well lubricated and covered with either clear PVC film or the cut-off digit from a glove. Other options are to use the probe without removing the adhesive backing or to carefully trim, using scissors, the adhesive off the pulse probe and than to wrap Koban around the palm of one hand and around the probe. Figure 3 presents bullae formation due to pulse oximetry probe.

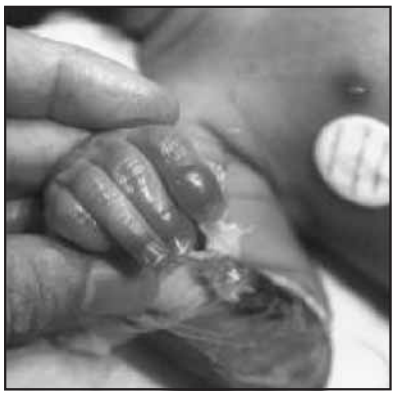

Figure 3. Blistering beneath the pulse oximetry probe

Despite an obvious susceptibility to cause blisters beneath the cuff, non-invasive arterial pressure monitoring can be used. Automated devices have potentials for the generation of high cuff pressures, and for this reason, sphygmomanometers measuring the arterial pressure are used. The skin under the sphygmomanometer cuff should be covered with PVC film and padding (wrap). Electrocardiogram monitoring is difficult because the adhesive pads can cause trauma on application or removal. For the ECG, all the skin sites are covered with either paraffin gauze/gelpad before self-adhesive electrodes are applied. The adhesive part can also be removed and the electrode laid beneath the patient or wrapped in mummified thoracic dressing to obtain good contact and a reasonable trace.

Intra-arterial monitoring has been recommended previously as the measurement of choice. However, occlusive pressure required to stop bleeding on removal of the cannula may cause an unnecessary trauma.

Induction of anesthesia should be as struggle free as possible. If there is an intravenous cannula or veins that look easy to cannulate, an intravenous induction with appropriate agent is preferred. Thiopentone has been used successfully in many patients with no adverse effects. Inhalation induction is also acceptable, especially with the advent of sevoflurane, and may be preferable in those patients with difficult venous access. For the maintenance of anesthesia, conventional methods of balanced anesthesia are suitable. Suxamethonium and nondepolarizing muscle relaxants are frequently used in EB patients without adverse effects.

Probably the most important, but problematic aspect of anesthetic management in PA-JEB patients is maintaining a clear and secure airway (9). The first issue beyond safely maintaining an airway is avoidance of further bullae. Traditional manipulation of the airway is difficult because a face mask cannot be applied firmly. More gauze must be placed beneath the anesthesiologist's fingers while doing a minimal chin lift. Head tilt can be achieved by gentle manipulation of the head with a hand beneath the occiput. Intubation was advocated to avoid the trauma related to the face mask use. Gloves, laryngoscope blades, airways etc., should also be well lubricated. To reduce the risk of new laryngeal bullae formation, a tracheal tube (TT) a half to one size smaller than predicted may be necessary: and if a cuffed tube is required, then the cuff must be carefully inflated. The tube should be secured (properly tied) with either ribbon gauze or Vaseline gauze and a 'collar' of adhesive tape around the TT to prevent the ties from slipping.

At the end of the operation, pharyngeal suctioning should only be performed under direct supervision, taking care not to place the sucker tip the on the mucosa directly. Prior to extubation, the pharynx and larynx should also be inspected for new bullae formation (10).

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ANESTEZIJA KOD NEONATUSA SA SINDROMOM ATRESIO PYLOR I JUNCTIONAL EPIDERMOLYSIS BULLOSA
Prikaz slučaja

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SAŽETAK

U radu je prikazan opis anesteziooloških postupaka kod neonatusa sa atresio pylori-junctional epidermolysis bullosa (PA-JEB) sindromom. Neonatus sa PA-JEB sindromom može predstavljati anesteziološki izazov čak i za najiskusnijeg anesteziologa pri čemu uvek treba imati u vidu nekoliko osnovnih principa koji omogućavaju izvođenje neophodnih postupaka.

Bilo kakvo tangencijalno povlačenje kože uslovljava nastanak buloznih formacija za razliku od sila pritiska koje se uglavnom tolerišu. Athezivni flaster i trake, athezivne ECG elektrode kao i athezivni nastavci za pulsnu oksimetriju ne smeju biti korišćeni ni pod kakvim uslovima. Površinu tela neonatusa koja će biti izložena manipulaciji, povlačenju i pritiscima treba bogato obložiti vatom. Pri inhalacionom uvodu maske treba premazati sredstvom za omekšavanje ili višeslojno obložiti vazelinskim gazama. Laringoskop i orofaringealni tubus moraju biti dobro podmazani lubrikantom rastvorljivim u vodi – ne koristiti Lidokain gel. Treba izbegavati faringealnu aspiraciju. Ne postoje kontraindikacije za trahealnu intubaciju pri čemu treba voditi računa o veličini endotrahealnog tubusa.

Uz maksimalnu zaštitu kože i sluznica moguće je izvesti anesteziju kod dece sa PA-JEB sindromom uz minimalne sekvele.

Ključne reči: **PA-JEB sindrom, neonatus, anestezija**