



Urological surgery in epidermolysis bullosa: tactical planning for surgery and anesthesia

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ABSTRACT

Epidermolysis bullosa (EB) is characterized by extreme fragility of the skin and mucosae. Anesthetic and surgical techniques have to be adapted to those children and routine practice may not be adequate. Urological problems are relatively common, but surgical techniques adapted to those children have not been well debated and only low evidence is available to this moment. Herein we discuss the specifics of anesthetic and surgical techniques chosen to treat a six year old EB male presenting with symptomatic phimosis.

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INTRODUCTION

There are four different types of hereditary epidermolysis bullosa (EB), with an incidence of almost 20/1000000 live births, all of them characterized by the extreme fragility of the skin and mucosae (1). Minimal trauma can lead to blistering and/or ulcerations that may complicate with secondary infections and scarring. Blistering may affect esophageal, tracheal, urethral, bladder and ocular surfaces, causing dysphagia, esophageal, distal ureteral and/or urethral stenosis, dysuria, voiding dysfunction, respiratory and ophthalmological problems.

Urological complications from EB predominate in junctional EB (1). Common urological conditions may also affect those children. Phimosis, specifically, is commoner in EB patients than in normal children, due to repetitive blistering and scarring of the prepuce (1).

Anesthesia in EB children is complicated by the difficulties involved in skin and mucosal manipulation. Airway instrumentation may lead to complications. Monitoring may be difficult, due to difficulties to fixate any devices to the skin. Special tactics and careful preoperative planning for surgery and anesthesia in EB children are needed.

DESCRIPTION

KOS, a six year old male, presented to the Pediatric Urology Clinics of Antonio Pedro University Hospital for treatment of phimosis, irresponsive to topical steroids and complicated by episodes of dysuria. The boy was diagnosed with dystrophic EB as a neonate and has been treated by multiple subspecialists for complications

of the disease (lagophthalmos, trichiasis, bilateral corneal opacifications, pseudo-syndactily affecting hands and feet, recurrent oral ulcerations, undernourishment, failure to thrive, constipation, dental caries and periodontitis). Considering the healthy aspect of penile skin except for the phimotic ring and the age of the child, elective surgery under general anesthesia was scheduled. The urinary meatus was normal (Figure-1).

Figure 1 - Right and left foot (A and B, respectively), right and left hand (C and D, respectively), lagophthalmos and corneal opacifications, dermatologic problems over the nose, where the mask was applied (E) and phimosis/penile skin (F).



After preliminary sedation with oral midazolam the patient was anesthetized by a mixture of sevoflurane and propofol through a facial mask complemented by a penile block. Airway manipulation was avoided by the exclusive usage of silicone invested facial masks. Two different sizes of masks were alternated each 15 minutes in order to avoid continuous trauma to the skin. A peripheral vein was punctured in the right foot, after application of dosed manual proximal compression of the leg, after segmental involvement with soft cotton bandages ("human tourniquet"). After puncture the skin the foot was protected with a layer of orthopedic soft cotton and an elastic bandage, to which the venous catheter was fixed by a knot and adhesive tape, without direct fixations of the catheter to the skin. Prophylactic antibiotics were administered (cefazolin, 20mg/kg). The electrocautery was grounded by a metal plate tied to the skin using a soft bandage. Cardiac activity was monitored by three baby-sized adherent electrodes, previously cut to minimize the adherent surface and were later removed with extreme care. Peripheral oxygen saturation was monitored by a clip fixed to the right foot. Folds or irregularities in bed clothing were carefully avoided.

The stenotic distal preputial ring was carefully dilated to expose the glans. There were no blisters and the urinary meatus was normal (Figure-2). A shortened frenulum was divided. The preputial ring was caught by a pair of forceps and submitted to distal traction. The surgeon marked the ideal place for skin sectioning on the external preputial surface. A clamp was inserted distal to the skin mark, saving the remaining penile skin from manipulation (non-touch technique). The distal stenosed preputial ring was then resected. Cutaneo-mucosal suture was done aided by four stay sutures, with a minimum-touch technique, avoiding skin manipulation with forceps, with separated intradermal stitches of 6.0 vicryl®. The suture, distal skin and glans were covered with dexametasone cream. Despite maximum care, at the end of the procedure the patient showed a small area of blistering in the proximal dorsal penile skin (Figure-2).

Post-operatively the patient received oral analgesia exclusively with paracetamol. Anti-

biotics (cephalexin) were used for 5 days, with chlorhexidine baths three times a day and topical therapy with dexametasone cream, also tid. The incision was completely healed after 1 week with good esthetic and functional results. The mother was counseled to avoid meatal trauma and to maintain constant protection of the glans/meatus with vaseline ointment. Clinical and ultrasonographic urological follow-up each six months was offered, in order to detect eventual long-term complications.

DISCUSSION

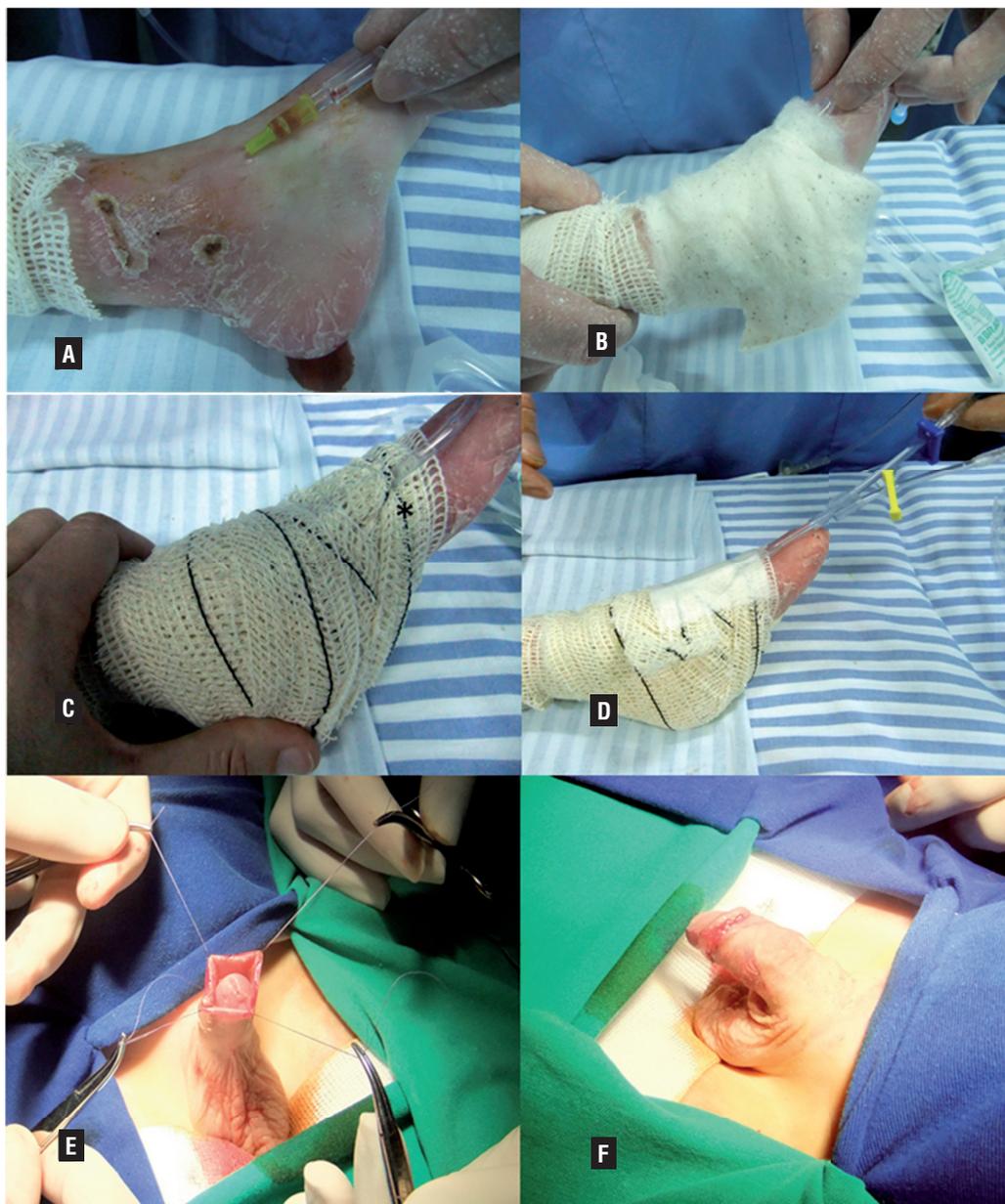
EB types are defined by histology, genetic analysis and clinical phenotype. Simplex EB affects only the basal layer of the skin. The blisters heal easily and predominate in hands and feet. Severe forms are represented by junctional EB, dystrophic EB and Kindler disease. Those three forms of the disease affect the skin and mucosae and have a bad prognosis: early death is common and sequelae are universally present. The main causes of death are infections (in children), aggressive skin tumors and kidney failure (in young adults). Treatment is supportive, despite recent trials of genetic therapy, bone marrow transplantation and fibroblast injections.

The establishment of the American EB Registry revealed a 17 to 30% incidence of urological complications, mainly in junctional and dystrophic EB (1), mostly affecting males (2:1), probably due to urethral anatomical differences. Their incidence grows proportionally to the patients' age. Even so, perhaps due to the rarity and bad prognosis of the disease, reports about urological complications of EB are rare. Long-term consequences of urological treatment are difficult to evaluate, as the available level of evidence is low and many patients die early from other EB complications. Blistering may affect the urethra and the bladder, but until this moment ureteral involvement has never been demonstrated. Meatal and ureterovesical junction stenosis (presumably due to scarring of the bladder internal surface) are the most frequent problems. Patients may present severe dysuria, frequency, urinary retention, UTIs (especially if extensive

groin skin lesions are present) and hematuria (2). In some cases secondary hydronephrosis and renal failure develop (3). Meatal stenoses are difficult to treat, as they are prone to recur after calibration and/or meatoplasties; intermittent catheterization is contraindicated (to avoid urethral trauma), perma-

nent catheters and urinary stomas are problematic because of peristomal dermatitis. Internal bypasses with double J catheters are the most frequent options to treat ureteral obstruction nowadays (2), despite ureterosigmoidostomies being sporadically used (3). Considering the risks of silent urological

Figure 2 - Vein catheter fixation: A peripheral vein was punctured in the right foot (A). The skin was protected by a cotton bandage (B). The cotton bandage was covered by an elastic bandage and the venous catheter was fixed to the bandage by a knot (*) (C). The catheter was fixed again to the bandage by an adhesive (no adhesive tapes were fixed to the skin) (D). An effort was done not to touch the penile skin to insert the stitches between the stay sutures (non-touch technique) (E). Final result (F).



complications, especially obstructive hydronephrosis and the low morbidity of the exam, we propose that those patients should be periodically evaluated by ultrasound and followed up by urologists. As a rule, urinary tract instrumentation should be avoided. Classical urodynamics, cystourethrograms and cystoscopy should be avoided when possible. If needed, urethral catheterization should be done with atraumatic low caliber catheters after very careful urethral lubrication with the child's cooperation or sedation. In girls labial sinequiae may develop (4). Sexual activity is especially problematic, considering the potential for genital trauma, and is an important clinical problem after adolescence. Only patients affected by the milder forms of the disease are able to participate in intercourse.

Despite almost 40% of the affected males in the USA being circumcised, 50% as neonates (1), we were unable to find any details about the ideal surgical technique in the literature. There are no reports about suture technique or eventual usage of clamps, especially the commonly employed Plast-bell clamp. Surgery should be avoided in the presence of penile blisters. No problems related to poor/delayed healing or abnormal scarring has been reported. Cases of symptomatic phimosis developing in adulthood, including some cases of acute urinary retention have been presented (1). A relationship between circumcision and meatal stenosis has been suggested by Cherif (5) and the constant exposure of the meatus to friction against the underwear could potentially predispose to meatal stenosis, perhaps advocating against routine circumcision in EB boys. However, circumcision seems not to be related to urethral stenosis in the American EB project cohort and symptomatic phimosis must be treated also in this population. Comparative studies focusing circumcised and uncircumcised EB populations are not available. Circumcision per se may be related to meatal stenosis, independently of any other factors: some authors suggest a 20% incidence of meatal stenosis after routine neonatal circumcision in healthy boys (6).

Considering the absence of established surgical protocols, we opted for classical circumcision with a no-touch technique, to spare the re-

maintaining skin from trauma by using instruments only in the skin previously selected to be resected and putting stay sutures to expose the incised surfaces without the need to use forceps to place the remaining stitches (Figure-2). Reabsorbable intradermal stitches were chosen, in order to minimize postoperative skin trauma. A circumcision device (such as a plast-bell clamp), despite minimizing surgical time and manipulation, was judged unwise, as it could possibly induce contact glans ulceration during the period of retention. We opted for a brief period of prophylactic oral antibiotics (5 post-operative days) and topical steroid cream over the glans and distal prepuce, in order to avoid secondary infection and to minimize local inflammation during the early post-operative period.

Frailty of the skin, iron deficiency anemia, limited oral opening, poor dentition, adhesions of the tongue, eye problems and pseudosyndactily have to be considered in anesthesia planning (7). Airway complications are common and may lead to acute respiratory insufficiency or prolonged intubation. Laryngeal or tracheal manipulations should be avoided whenever possible and tracheal tubes should be well lubricated, half to one size smaller than the normal caliber to the age and with a low pressure cuff (8). Local anesthesia, despite eluding airway manipulation, is not practical in a non-cooperative child. Physical restriction is not an option, as restraining a struggling child leads to new cutaneous lesions. A pre-anesthetic sedative is useful, but rectal medications are contraindicated, to avoid anal trauma. For small procedures the maintenance of general anesthesia exclusively by facial masks is possible without any undue risks. The mask should be soft and unnecessary trauma to the face should be avoided. Other authors have described face mask + penile block for circumcision in an EB boy (9). We propose also to alternate different mask sizes from time to time, in order to vary the pressure sites during the operation, which is a newly proposed attitude. Other authors suggest lubricating the contact surface of the mask, but that may disturb the maintenance of the right position of the device. A key knowledge is that attrition or appliance of shearing forces to the patient's skin

are critical, but direct pressure does not normally cause new bullae. Cutaneous fixation of any device should also be avoided. After the puncture the skin was protected with bandages to which the catheter was fixed. Other authors suggest suturing the catheter to the skin (10) or the usage of special silicone tapes that, unfortunately, are not commonly available. Adhesive grounding pads to the electrocautery are contraindicated. Grounding may be obtained by using a metal plate or the new dry pads. Electrodes for cardiac monitoring should be used in minimal number and smallest size available. Some have dismissed any fixation of the electrodes, just maintaining them on the back of the supine patient and using the motionless patient's weight to maintain position, with or without added fixing silicone bandages (11). Needle electrodes are also an option for anesthetized children. We opted to use a minimal amount of adhesive to fix three electrodes, with extreme care to take them away after surgery, and were able not to cause new bullae, as observed in the first week follow-up.

CONCLUSIONS

EB is frequently related to urological complications (17 to 30%), with a very high morbidity, but surgical techniques adapted to those patients are not well discussed and there is only a low level of evidence available. Anesthesia also needs extensive pre-operative planning. Surgical planning for those patients has to consider the need of atraumatic techniques and to avoid any post-operative skin trauma by dressings, stitches or surgical devices. We propose herein that plas-tbell clamps may be inadequate for those children, which may be better treated by classical circumcision, with a non-touch technique.

CONFLICT OF INTEREST

None declared.

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