



Case Report

Developmental dysplasia of the hip in an epidermolysis bullosa patient; how to deal with surgical difficulties: A case report

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ABSTRACT

Epidermolysis bullosa (EB) is defined as a separation of the dermoepidermal junctions resulting in an increased risk of infection and the formation of fibrous tissue. We are presenting a case of a 22-month-old girl with developmental dysplasia of the left hip and EB. The patient was operated on, with the left hip open reduction, acetabuloplasty and hip spica application. For a safe surgery on EB patients, pre-operative anesthesia assessment is necessary to ensure proper neck movement and examine the mouth and airway for the presence of blisters. Certain intraoperative measures should be taken, like gentle handling of the patient to reduce the events of blistering and scarring. Avoiding all adherent dressings, tapes, and stickers, using non-adhesive dressings and applying a good cotton padding for patients, are crucial steps in such cases to achieve good results. Surgery for this patient went smoothly without acute complications. In this report, we hope to provide helpful information on the surgical management of such patients.

Keywords: Complications, Developmental dysplasia of the hip, Epidermolysis bullosa, Perioperative, Surgery

INTRODUCTION

Developmental dysplasia of the hip (DDH) describes a spectrum of structural abnormalities of the hip joint, where there is an abnormal relationship between the femur head and the acetabulum. Its incidence is 1–34 cases per 1000 live births. The treatment depends on the age of the child on presentation. Prompt diagnosis and early treatment when DDH is suspected are essential to have the best outcome possible.^[1] Treatment options include Pavlik harness in the first 6 months,^[2] and a closed reduction with hip spica from 6 to 18 months,^[3] and an open reduction for patients from 18 months to 8 years, in older adolescents and adults, other pelvic osteotomies and total hip arthroplasty may be useful.^[4] One old demographic study of DDH, in Saudi Arabia, showed an incidence rate to be found around 3.5/1000 live births, making DDH a rather familiar condition in Saudi Arabia.^[5]

Epidermolysis bullosa (EB) is a genetic disorder defined by the separation of the dermoepidermal junctions, leading to the formation of skin blisters resulting from minor mechanical trauma to the

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skin. This results in an increased risk of infection, the formation of fibrous tissue, and subsequent contracture and deformity.^[6]

According to the degree of tissue detachment within the basement membrane zone, EB has been categorized into three main types, EB simplex (EBS), junctional EB (JEB), and dystrophic EB.^[7] Clinical presentation differs with each subtype. It ranges from localized manifestations such as esophageal strictures and pseudosyndactyly to life-threatening conditions including recurrent skin infection and skin cancer. Management is mainly symptomatic with the treatment in an experimental stage.^[8]

A study from the eastern province of Saudi Arabia by Abahussein *et al.* found 16 cases over 7 years (1984–1990), which demonstrates the rarity of EB in the country. With JEB being the most prevalent type.^[9] Patients with EB are a high-risk group for anesthesia due to malnutrition, anemia, hypoproteinemia, dilated cardiomyopathy, and limited mouth opening. Children with EB are more vulnerable and less tolerant. Minor and trivial operations such as dressing changes, body mapping, and formation of hand molds are sometimes performed under general anesthesia.^[10]

Wounds in EB patients harbor diverse microbes. In a retrospective analysis of 739 wound cultures from 158 patients, 152 patients had positive culture, *Staphylococcus aureus* (SA) was found in 131 patients (86%), followed by *Pseudomonas aeruginosa* (PA) from 56 (37%) and *Streptococcus pyogenes* (GAS) from 34 (22%). Sixty-eight percent of the patients had cultures positive for methicillin-sensitive SA, and 47% had methicillin-resistant SA.^[11] This paper highlights the surgical difficulties faced by pediatric orthopedic surgeon in this challenging case and how to tackle them.

CASE REPORT

A 22-month-old girl was referred from a peripheral hospital as a case of the left DDH and an unknown skin condition for further evaluation and treatment. She is a product of a full-term pregnancy that was delivered vaginally. This patient was evaluated by the orthopedic service initially. The family reported no other medical history or complaints, and there were no allergies or significant family history.

The patient was looking well with no distress or cyanosis. However, there were blisters all over the body and on the hands and feet with whitish scars and thick fingernails. Hip examination revealed multiple diffuse bullae with erosions covered by hemorrhagic crust.

There was no tenderness or hotness, and the range of motion was limited, specifically in abduction. Galeazzi's test was positive, and the left lower limb was shorter than the right by 1.5 cm. Her pelvic radiograph [Figure 1] showed a left hip DDH.

The patient was referred to dermatology and genetics for further evaluation and treatment of the skin condition. Dermatology's impression was that the patient has EB (simplex vs. dystrophic subtype). They planned to do a skin biopsy for electron microscopy and histopathology. In addition, they advised routine wound care nursing and wound care education, frequent potassium permanganate bath, keeping the skin moist and avoiding dryness, application of hydrocortisone ointment, and the use of soft dressings. Skin biopsy confirmed the diagnosis of EBS type. There was no objection to surgery but only precautions while manipulating the patient. Genetics have seen the patient in their clinic, confirmed the diagnosis, and counseled the family.

When the patient became 2 years old, the patient was admitted for the left hip open reduction, acetabuloplasty, and hip spica application. Careful pre-operative anesthesia assessment was done to ensure no limitation in neck movement or mouth or airway blisters. Wound cultures were taken from both axilla and groin and were negative for MRSA. Hence, standard antibiotic prophylaxis, Cefazolin 30 mg/kg, was given in IV form. Intraoperative measures included gentle handling of the patient to reduce the events of blistering and scarring. They also included avoiding all adherent dressings, tapes, and stickers, using non-adhesive dressings and electrocardiogram electrode pads and applying good cotton padding for the patient. The surgery went smoothly without acute complications [Figure 2].

For preparation, iodine was used cautiously in small amounts, and a sterile, non-adhesive dressing was applied to the skin for draping to prevent skin injury when removing the drapes at the end of surgery. There was no skin blistering or reaction after these measures were taken.



Figure 1: Pelvic radiograph, shows a left hip developmental dysplasia of the hip.

An anterior hip approach, with an interval between the sartorius and the tensor fascia latae, was utilized. Deep dissection was carried out; then, the capsule was incised in a T-shaped fashion. The femoral head was reduced and capsulorrhaphy was performed that was followed by Pemberton acetabuloplasty. A K-wire was left going from the greater trochanter to the pelvis. Although it is not the standard of care to leave the K-wire in place after surgery, we appreciated that the patient had some ligamentous laxity as well as the lack of reports guiding the management of DDH in EB patients directed us that way.

She was discharged the next day after evaluation. A near follow-up was not possible as the family was from the northern part of the country and did not prefer a near follow-up. Hence, the patient was followed up on a regular basis.

The patient was admitted again after 1 month for K-wire removal, hip spica change, and re-examining the skin condition, which did not show any acute complications.

Intraoperatively, the skin did not have any complications [Figures 3-5]. After 6 weeks, the hip spica was removed

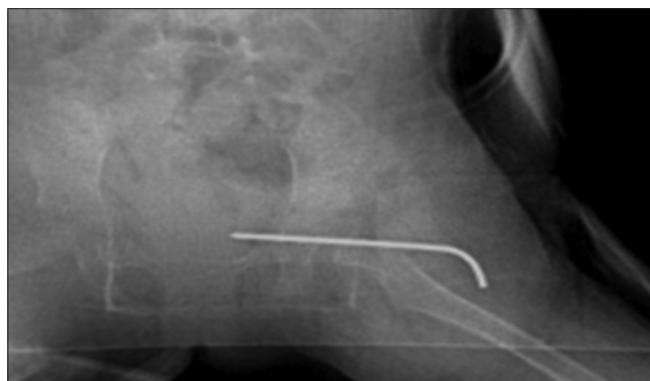


Figure 2: Post left hip open reduction and acetabuloplasty radiograph.



Figure 3: Intraoperative images of the skin (second surgery).

without complications. Following the hip spica removal, the patient was followed in the clinic virtually through the phone due to the COVID-19 pandemic and the family reported no complications. The patient has resumed follow-up with the regular clinic now that the pandemic is over. The patient then presented to our outpatient clinic for a follow-up visit 6 months post-operative, and she was 3 years old. She was able to walk with assistance. Hence, we referred her to physiotherapy for gait training. The patient was given a follow-up visit 6 months later, where she was doing fine with no new complaints and normal growth parameters. However, she was facing some difficulties when walking without assistance. Follow-up imaging showed acceptable hip reduction and preserved Shenton's line. Although a good reduction was achieved, the femoral head showed signs of avascular necrosis [Figure 6].

DISCUSSION

The involvement of the groin and the lower limb skin in this patient imposed serious difficulty in the casting and manipulation. Therefore, a thorough musculoskeletal assessment must be done to look for extensive contracture and osteopenia or osteoporosis, which may result in difficulties in achieving optimum procedural positioning. When splinting and casting are performed, it is important to maintain clean surroundings due to the increased risk of infection. Furthermore, it is advisable to use a non-adherent dressing as the first layer of contact with the skin, such as a silicone non-adhesive dressing. When handling the patient, avoid rubbing or stroking the skin as friction might result in more blistering of the skin, as much as possible to reduce the possibility of blistering. When padding is used, provide extra padding, and strapping, providing extra protection to the skin.^[12] To prevent unnecessary friction to the skin, dressings should be left in place when feasible. If removing



Figure 4: Intraoperative images of the skin (second surgery).



Figure 5: Intraoperative images of the skin (second surgery).

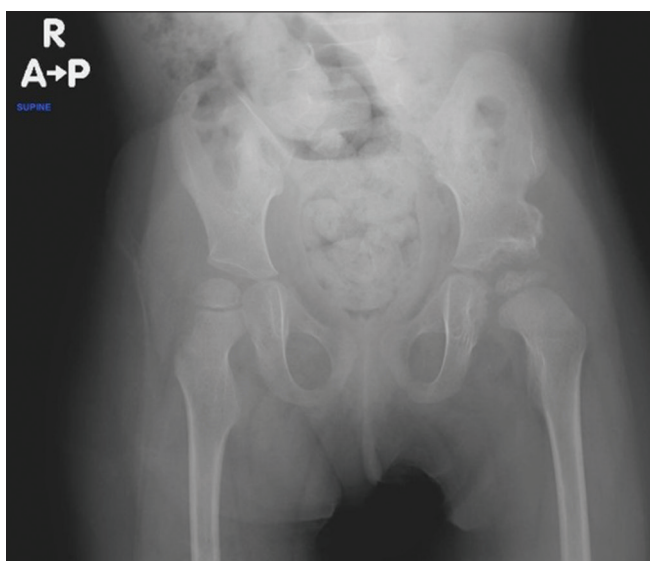


Figure 6: Follow-up radiograph, showing acceptable hip reduction and intact shenton line.

dressings is a must, a cling film can be used as a temporary measure, covering the skin. Suitable dressings must be used to avoid adhering to the skin and subsequent skin injury and ensure the safe removal of any dressing tape or monitoring stickers that may be inadvertently applied. The involvement of the skin and mucous membranes in the face, oropharynx, and neck represents a substantial challenge in airway management. Therefore, an extensive airway assessment must be done preoperatively to look for microstomia and limited mouth opening, fixed and scarred tongue, limited neck movement, poor dentition, and oral blistering are all common features, and should be identified preoperatively. Intraoperatively, the use of gentle pressure to distend the veins and aid cannula insertion to gain IV access without the need for tourniquet. If a tourniquet is used, it

should be well padded to avoid injuring the underlying skin. Equipments applied to the skin or mucous membranes should be lubricated before application, which will help reduce the shearing forces. Monitoring during anesthesia imposes a difficult challenge and requires the removal of all adhesive parts of the monitoring devices. It is also advised to use the clip-on type of pulse oximetry.^[13] When monitoring blood pressure, apply 2-3 layers of soft padding beneath the cuff to protect the underlying skin from friction and rubbing. As for electrocardiogram, the use non-adhesive electrode pads wherever possible.^[13-15]

There are some factors to take into account for post-operative wound management in EB patients. It is crucial to maintain hemoglobin levels above 80 g/L. Anemia in EB patients is usually multifactorial, resulting from chronic blood loss that these patients exhibit, as well as iron deficiency anemia. Adequate skin care to prevent bleeding and iron supplementation are recommended. Malnutrition resulting from poor nutrition combined with increased calory demand in EB patients is another contributing factor to poor wound healing. Low protein levels are associated with poor granulation tissue formation and subsequent poor wound healing process. Albumin is a valid indicator of nutritional status; levels <2-3 g/dL are associated with poor wound outcomes. For patients with EB, it is important to gently cleanse wounds using low-toxicity solutions such as saline solution, water, or dermol 500 (containing benzalkonium chloride 0.1% and chlorhexidine hydrochloride 0.1%). It is also advisable to avoid the excessive use of cytotoxic solutions such as (povidone-iodine).^[16] Postoperatively, EB patients must receive adequate analgesia to prevent distress that might result in excessive movement against the bed or crib. It is also important to consider that opioids contribute to pruritus, which is common among EB patients. The oral route is the preferred route of administration. However, suppositories are not recommended as they may lead to anal trauma in these patients.^[14] Moreover, it is important to administer post-operative antibiotic prophylaxis in such patients. The current patient received three doses of Cefazolin, considering the fact that our patient was not colonized by MRSA after obtaining wound cultures from both axilla and groin. However, in MRSA cases, Vancomycin is the drug of choice.^[17] Although an acceptable reduction was achieved, the femoral head showed signs of avascular necrosis. This might be attributed to the delay in the presentation of this case.^[18] Despite the fact that the results are unpredictable for the dysplasia in this particular age in patients with EB, the benefit of the surgery outweighs the risk of infection and worsening of the skin condition. A multidisciplinary team approach to managing such a patient is crucial. Pre-operative preparation must be individualized, and special consideration must be given to the potential anesthetic problems. When procedures under anesthesia are planned or are inevitable, it is recommended

to coordinate as many surgical procedures as possible to avoid unnecessarily repeated anesthesia.

CONCLUSION

Orthopedic surgeries performed on EB patients are safe if precautionary measures are applied correctly. Great care must be taken for EB patients preoperatively, intraoperatively, and postoperatively. Care must be taken and gentle handling of the patient to prevent injury to the skin, reducing the risk of blisters as well as infection must be considered.

AUTHORS' CONTRIBUTIONS

MAA and AHA conceived the idea, conducted literature research, provided research materials, and collected data, as well as initial and final manuscript preparation and editing. AKA, RMA, AIS, and AAA have prepared, edited, and provided logistic support. All authors have critically reviewed and approved the final draft and are responsible for the manuscript's content and similarity index.

DECLARATION OF PATIENT CONSENT

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient's parents have given their consent for the patient's images and other clinical information to be reported in the journal. The parents understand that the patient's name and initials will not be published, and due efforts will be made to conceal the patient's identity, but anonymity cannot be guaranteed.

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CONFLICT OF INTEREST

There are no conflicting relationships or activities.

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