RECESSIVE DYSTROPHIC EPIDERMOLYSIS BULLOSA IN A PREGNANT WOMAN. CASE REPORT.

López Torres MF [1], Ibargüengoitia Ochoa F [2], Lira Plascencia J [3], Canuto Hinojosa AM [4]

[1] *Obstetrics and Gynecology Resident at National Institute of Perinatology, Mexico City, Mexico.*
[2] *Obstetrics Department at National Institute of Perinatology, Mexico City, Mexico*
[3] *Adolescence Coordination at National Institute of Perinatology, Mexico City, Mexico.*
[4] *Anesthesia Department at National Institute of Perinatology, Mexico City, Mexico*

Objective: Bullous epidermolysis encompasses a group of diseases characterized by extreme fragility of the skin and mucous membranes, resulting in the formation of blisters after minimal trauma. The following case is reported as there is little literature on pregnancy in women with this condition. Special care must be taken during therapeutic interventions in order to avoid the formation of bullae or exacerbate those already present. The case is presented to highlight the implications of care both at the time of birth as well as anesthetic considerations.

Methods: We report a patient with recessive dystrophic epidermolysis bullosa and its resolution. 25-year-old patient, originary and resident of a rural area of the state of Morelos, México; diagnosis of dystrophic epidermolysis bullosa at birth. She began prenatal care at 23 weeks of gestation with a body mass index of 16.1 m2/kg. At week 29, she was hospitalized for transfusion and infusion of iron salts given a hemoglobin result of 5.2 g/dl; At week 35 she already had hemoglobin of 11.5 g/dl. At 34/5 weeks, the diagnosis of intrauterine growth restriction was made. In week 38, the pregnancy was electively resolved through elective surgery under regional anesthesia. A live newborn was obtained weighing 2490 grams and an Apgar score of 9-9 at minute one and five respectively. The baby did not present any manifestations of the disease. The patient and her baby were discharged on the third post-operative day in good condition.

Results: Women with epidermolysis bullosa are not at increased risk for complications associated with pregnancy.

Antenatal care. - Genetic counseling should be offered to patients given their hereditary pattern. Malnutrition, severe anemia and chronic infection can be associated; In cases with hemoglobin less than 8 g/dl, transfusion and/or intravenous iron should be considered [1].

Intrapartum considerations. - Although there are no differences between the outcome of vaginal birth and cesarean section, the risks and benefits should be discussed in all cases. Peripheral venous access should avoid excessive tightening of the tourniquet and secure the catheter with silicone tape and protect with padding and non-compressive bandage.

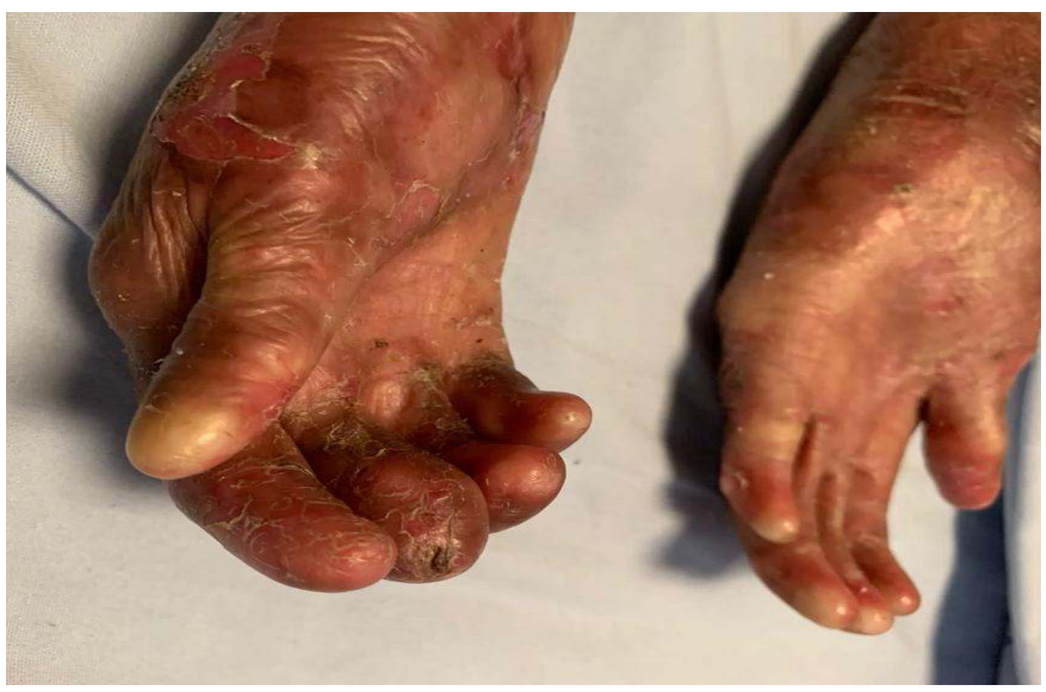

Figure 1. Pseudosyndactyly and contractures

The operating table must have a soft and padded mattress, cut the adhesive from the electrocautery plate and only place the conductive medium with a sufficient amount of gel in contact with the skin, use surgical drapes without adhesives, consider a wide surgical incision in the case of a cesarean section, subcuticular sutures can be used to suture the skin, avoid vigorous stimulation of the baby at the time of birth, avoid continuous oximetry and use a silicon Foley catheter [2].

Regional anesthesia. - It seems to be the most favorable method in patients with epidermolysis bullosa in order to avoid airway manipulation [3,4].

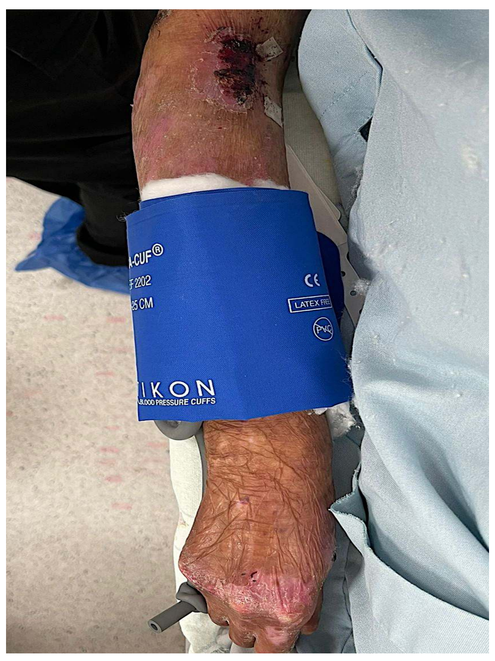

Figure 2. Cuff with padding

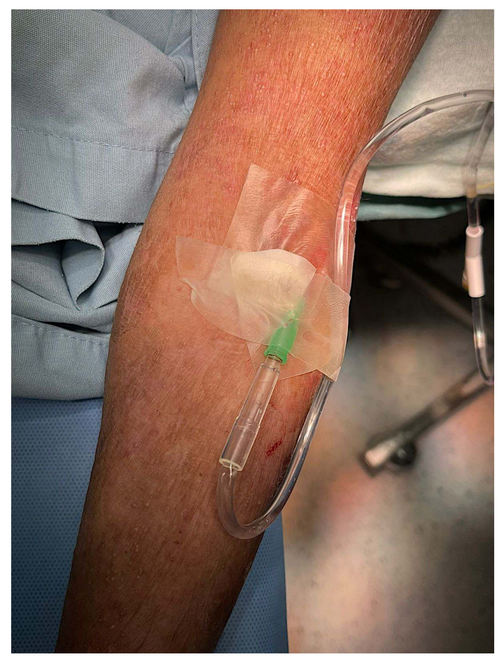

Figure 3. Adhesive-free venous access

Intrapartum anesthesia. - It is recommended that the patient position herself. For monitoring it is recommended to gently take blood pressure and oximetry as well as space measurements [5].

Postpartum considerations. - Compression stockings should not be used due to the force produced by their application. Intramuscular injections can be administered if necessary, avoiding areas of injury. Patients may require extra help caring for their baby, especially if the mother's hands are affected by blisters, scars, or pseudosyndactyly.

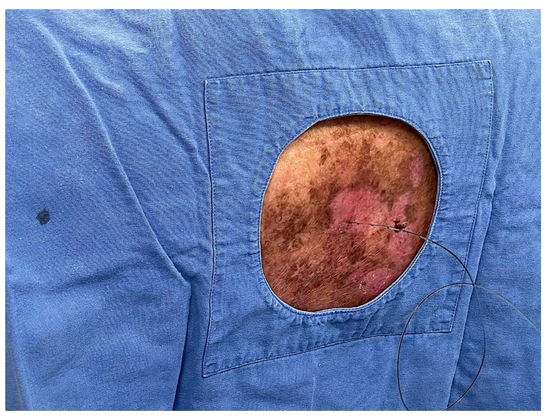

Figure 4. Catheter in an area without bullae

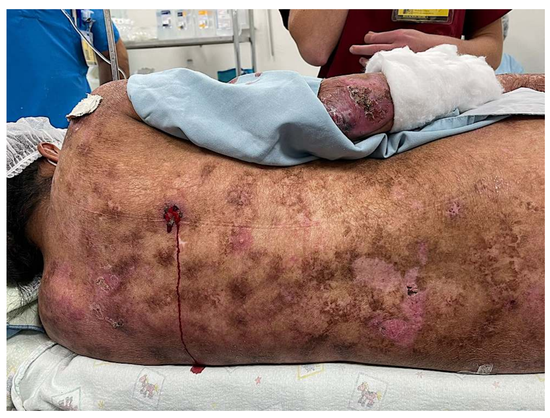

Figure 5. Patient´s back before regional anesthesia

Breastfeeding. - It is not contraindicated. A well-lubricated silicone nipple shield should be offered.

Conclusion: Epidermolysis bullosa is a serious, disabling skin condition. Patients should be informed that pregnancy is not a contraindication but that, like any woman, they should be in optimal health and have good nutritional status before attempting pregnancy. Importantly, there do not appear to be any complications in these pregnancies and the skin condition does not appear to worsen as a result of the pregnancy. The most critical moment is the one related to birth. Regional anesthesia should be recommended for minimal impact on the puncture site. Pregnancy and birth in patients with epidermolysis bullosa can be managed safely, but it is important to plan all situations by a well-informed multidisciplinary team.

References

1. Simpson B, Tarango C, Lucky A W. Clinical algorithm to manage anemia in epidermolysis bullosa. Pediatr Dermatol 2018; 355(5):1-2. doi:10.111/pde.13587.
2. Shah N, Kumaraswami S, Mushi J E. Management of epidermolysis bullosa simplex in pregnancy: a case report. Case Report Women`s Health 2019; 24: e00140. doi: org/10.1016/j.crwh.2019.e00140.
3. Bolt LA, O'Sullivan G, Rajasingham D, Shennan A. A review of the obstetric management of patients with epidermolysis bullosa. Obstet Med [Internet]. 2010;3(3):101–5. Disponible en: http://dx.doi.org/10.1258/om.2010.100009
4. Araújo M, Brás R, Frada R, Guedes-Martins L, Lemos P. Caesarean delivery in a pregnant woman with epidermolysis bullosa: anesthetic challenges. Int J Obstet Anesth [Internet]. 2017;30:68–72. Disponible en: http://dx.doi.org/10.1016/j.ijoa.2017.01.010
5. Scherhag A, Dick W. Special aspects of anesthesia in patients with epidermolysis bullosa based on a case example. Anaesthesiol Reanimat 1998; 23(5): 129-33. PMID 9854331.