



LABOR IN PREGNANT PATIENT WITH EPIDERMOLYSIS BULLOSA

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BACKGROUND

Epidermolysis Bullosa (EB) is a disorder caused by genetic mutations of structural proteins in the basement membrane causing abnormal adhesion leading to fragile skin. Friction and trauma to the skin can lead to bullae and blisters. EB has an incidence of 1 in 50,000 births and can affect multiple organ systems including dermatologic, cardiac, gastrointestinal, and musculoskeletal. (1)

There are four main types of Epidermolysis Bullosa: EB Simplex, Junctional EB, Dystrophic EB, and Kindler EB

- EB Simplex: Autosomal dominant inheritance characterized by blistering in the epidermis. (1)
 - Skin blistering with ulcerations
- Junctional EB: Autosomal recessive inheritance characterized by skin blistering in the lamina lucinda. (1)
 - Ulcerations, **dental involvement, and absence of nails**. Can also be associated with **tracheolaryngeal stenosis**, dilated cardiomyopathy, and anemia
- Dystrophic EB: Autosomal dominant or recessive inheritance characterized by skin blistering beyond the basal membrane layer (1)
 - Ulcerations, **hyperkeratosis of hands and feet, pseudo-syndactyly**, dilated cardiomyopathy, **esophageal stricture**.
- Kindler EB: Autosomal recessive inheritance characterized by blistering in the basement membrane and/or lower (1)
 - Skin atrophy, **poikiloderma**, gum erosions, and GI/GU involvement.



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CASE REPORT

21yo G1P0 @ 36w6d presented to labor and delivery unit for scheduled induction of labor.

- Her risk factors include recessive dystrophic EB with associated dental erosion, small mouth opening, diffuse bullae, hyperkeratosis of hands and feet, pseudo-syndactyly
- She would be a high-risk cesarean section, if necessary, due to her skin fragility secondary to EB, concerning airway exam, and unknown possible airway strictures that are associated with dystrophic EB.

Anesthetic plan include ultrasound guided IV, early epidural, and emergency airway equipment prepared.

- Dressing plan was discussed with wound care team.
- Anesthesia resident placed ultrasound guided IV with careful awareness to not apply any shear pressure on skin even with site prep or ultrasound movement. Tourniquet was placed over gown. Dressed with Mepitel (silicone based mesh).
- Epidural placed with the following considerations:
 - Betadine solution dabbed on for site preparation. No chloraprep
 - Assured epidural site had no bullae or skin blister. Only one pass with lidocaine with no skin wheal.
 - Epidural secured with Mepitel and additionally secured to patient's gown with tape
 - Level was frequently checked throughout labor
- Nurses in mother baby unit were given adhesion removal spray for when epidural and IV needed to be removed

Patient had successful uncomplicated vaginal delivery. 2nd degree laceration noted and repaired by OB with no complications. She was discharged postop day 2. No skin-on-skin contact with mother and baby due to EB.



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DISCUSSION / TEACHING POINTS

This case reveals the complexity of EB. It highlights the need for meticulous multidisciplinary planning with regards to patient's comfort and safety.

- Concerns include not worsening patient's dermatologic pathology, assessing airway plan in case of epidural analgesia failure, and postop plan when patient is off L&D unit
- Safe methods for obtaining patient's vitals including clip on pulse oximeters, layer of cotton wool padding and gown under blood pressure cuff.

Anesthetic plan included:

- Early epidural to allow time to prepare and troubleshoot
- OR preparation including small endotracheal tube, McGrath, fiberoptic tower, and premade silicone strips for EKG stickers
- Dressing plan discussed with wound care team, labor and delivery nurses, and postpartum mother baby nurses